Section 8

Discontinuing immunoglobulin replacement therapy

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In patients whom the immune defect is thought to be transient or an empirical trial of IRT is indicated prior to confirming the diagnosis, IRT can be discontinued, the patient monitored and IRT recommenced if clinically indicated. Ideally discontinuation should not coincide with time points when the risk for infections is increased. For accurate assessment of the efficacy during seasonal variation, IRT must only be discontinued after a minimum of 12 months. If IRT is for a limited time this must be discussed upfront, including counselling of the patient and family about the risks and benefits of cessation of therapy, clinical assessment and laboratory testing.

Reasons to discontinue IRT

7.1 Class III or IV PID

After 12 months, stop IRT and evaluate immune responses six months after last infusion. If the improvement is sustained for 24 months, IRT may be discontinued. If infections recur, IRT must be recommenced, and the above process repeated. If infections recur thereafter, then lifelong IRT is required.[3-10] After PID curative treatment such as HSCT IRT can be discontinued after humoral reconstitution.[7]

7.1.2 Secondary immune deficiency states

If indicated, IRT should be replaced for 12-months, and the patient re-evaluated. IRT must be discontinued if a reduction in the frequency or severity of infections is not noted. If a longer duration of replacement is required, then annual assessments are recommended. [11-14]

7.2 Monitoring the patient after discontinuation

After discontinuation, the patient must be evaluated at four to six months, then at 12-months or as indicated in the event of recurrent infections, to assess the frequency and severity of infections and the presence of end organ damage.[16]

End organ damage includes features of bronchiectasis and cytopenias. As there is a good correlation between high resolution chest computer tomography (HRCT) scores and clinical findings, routine CT scan screening annually/biannually is not necessary.[17]

Laboratory investigations include IgG levels, antigen specific antibody vaccine responses[18-20] and a full blood count with white blood differential count . Ideal levels of serum IgG post-cessation of therapy have not been established and IgG results must rather be correlated with

the clinical history of frequency and severity of infections and should be above the lower limit of the age-related reference interval.[21]

Recommendations

1. IRT can be discontinued in patients with category III and IV PIDs[3-10], after PID curative treatment [7] and in patients with SID after a minimum 12-month trial [11-14]
2. Monitor the patient post-IRT discontinuation. This must include a clinical assessment, frequency and severity of infections, serum Ig levels and vaccine responses [16-21]
3. IRT must be recommenced if infections recur after discontinuation [16].

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