

BC Cancer Protocol Summary for Treatment of Cutaneous T-cell Lymphoma (Sézary syndrome) with Extracorporeal Photopheresis

Protocol Code

LYMFECF

Tumour Group

Lymphoma

Contact Physician

Dr. Kerry Savage

ELIGIBILITY:

- Patients with cutaneous T-cell lymphoma with erythrodermic mycosis fungoides with peripheral blood component and Sezary syndrome.

TESTS:

- Baseline (required before first treatment): CBC & diff, platelets, bilirubin, ALT, smear for Sézary cells, CD4 and CD8 counts, LDH, PTT, INR,
- Baseline (required, but results do not have to be available to proceed with first treatment; results must be checked before proceeding with further treatment): HBsAg, HBcoreAb
- Before each treatment: CBC and diff, platelets

PREMEDICATIONS:

None

SUPPORTIVE MEDICATIONS:

If HBsAg or HBcoreAb positive, start lamiVUDine 100 PO daily for the duration of ECP therapy and continue for one year from treatment completion for patients who are HBsAg positive and for six months for patients who are HBcoreAb positive.

TREATMENT:

Drug	Dose	BC Cancer Administration Guideline
Extracorporeal Photopheresis (ECP) methoxalen (UVADEX)	0.017 times the final buffy coat volume in millilitres (varies from 3-6 mL/treatment; 6 to 12 mg on two consecutive days every 4 weeks)	Infused into the product bag immediately before phototherapy

Reassess all sites of disease after 6 months. Initial treatment is 6 months. Consider a further 6 months of treatment for responders

DOSE MODIFICATIONS:

None

PRECAUTIONS:

1. **Photosensitivity:** Minimise exposure to sunlight and artificial UV light during treatment. It is recommended that patients wear sunscreen greater than or equal to SPF 15 and sunglasses for 24 hours after treatment.
2. **Hepatitis B Reactivation:** All lymphoma patients should be tested for both HBsAg and HBcoreAb. If either test is positive, such patients should be treated with lamivudine during chemotherapy and continue for one year from treatment completion for patients who are HBsAg positive and for six months for patients who are HBcoreAb positive. Such patients should also be monitored with frequent liver function tests and hepatitis B virus DNA at least every two months. If the hepatitis B virus DNA level rises during this monitoring, management should be reviewed with an appropriate specialist with experience managing hepatitis and consideration given to halting chemotherapy.

Call Dr. Kerry Savage or tumour group delegate at (604) 877-6000 or 1-800-663-3333 with any problems or questions regarding this treatment program.

References:

1. Alfred A, Taylor PC, Dignan F, et al. The role of extracorporeal photopheresis in the management of cutaneous T-cell lymphoma, graft-versus-host disease and organ transplant rejection: a consensus statement update from the UK Photopheresis Society. *Br J Haematol* 2017;177(2):287-310.
2. Willemze R, Hodak E, Zinzani PL, et al. Primary cutaneous lymphomas: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2013;24 Suppl 6:vi149-vi154.
3. NCCN Clinical Practice Guidelines for Primary Cutaneous Lymphomas Version 2.2019-Dec.17,2018
4. Edelson R, Berger C, Gasparro F, et al. Treatment of cutaneous T-cell lymphoma by extracorporeal photochemotherapy. Preliminary results. *N Engl J Med* 1987;316(6):297-303.
5. Cho A, Jantschitsch C, Knobler R. Extracorporeal photopheresis-an overview. *Front Med (Lausanne)*. 2018;5:236. Published 2018 Aug 27. doi:10.3389/fmed.2018.00236
6. Zic JA. Extracorporeal photopheresis in the treatment of mycosis fungoides and Sézary syndrome. *Dermatol Clin* 2015;33(4):765-76.
7. Zic J, Arzubiaga C, Salhany KE, et al. Extracorporeal photopheresis for the treatment of cutaneous T-cell lymphoma. *J Am Acad Dermatol* 1992;27(5 Pt 1):729-36.
8. Zic JA, Stricklin GP, Greer JP, et al. Long-term follow-up of patients with cutaneous T-cell lymphoma treated with extracorporeal photochemotherapy. *J Am Acad Dermatol* 1996;35(6):935-45.
9. Scarisbrick JJ, Prince HM, Vermeer MH, et al. Cutaneous Lymphoma International Consortium study of outcome in advanced stages of mycosis fungoides and Sézary syndrome: effect of specific prognostic markers on survival and development of a prognostic model. *J Clin Oncol* 2015;33(32):3766-3773.