



Hull University Teaching Hospitals and North Lincolnshire NHS Trusts
Haematology Multidisciplinary Team Guideline and Pathway

Peripheral T-cell Lymphomas (PTCL)

Cutaneous T-Cell Lymphoma will be referred to the Supranetwork Skin Lymphoma MDT (based at Leeds).

Full details on the Standard Operating Policy for The Supranetwork Cutaneous T-Cell Lymphoma MDT

https://www.wyhpartnership.co.uk/application/files/6415/0159/1518/TCell_SOP_2017_2_3.pdf

Background:

The Hull and North Lincolnshire Haematology Multidisciplinary team manages patients with haematological malignancies on three sites, Diana Princess of Wales Hospital Grimsby, the Queens Centre for Haematology and Oncology at Castle Hill Hospital Hull University Teaching Hospitals NHS Trust and Scunthorpe Hospital. Levels of service provided in these organisations is as defined in the NICE guidance “Haematological Cancers: improving outcomes NG47” 25th May 2016 <https://www.nice.org.uk/guidance/ng47/evidence/improving-outcomes-in-haematological-cancers-the-manual-pdf-2487893581>

Low-to-intermediate intensity chemotherapy is delivered in Grimsby, the Queens Centre Castle Hill Hospital and Scunthorpe Hospital. High-intensity chemotherapy and autologous stem cell transplantation is delivered at the Queens Centre, Castle Hill Hospital.

Guideline Details:

Peripheral T-cell lymphomas (PTCL) accounts for roughly 10% of all non-Hodgkin’s Lymphomas (NHL). The mature/peripheral T-cell neoplasms are a biologically and clinically heterogeneous group of rare disorders and in Europe peripheral T-cell lymphoma not otherwise specified (PTCL-NOS), anaplastic large cell lymphoma (ALCL) and angioimmunoblastic T-cell lymphoma (AITL) account for about three quarters of all cases.

Excluding anaplastic lymphoma kinase (ALK)-positive anaplastic large cell lymphoma (ALCL) and indolent mycosis fungoids, which have a good outcome, 5-year survival for other nodal and extranodal T-cell lymphomas is about 30%. Most patients present with unfavourable international prognostic index (IPI) scores (>3) and poor performance status (PS).

Diagnosis of peripheral T-Cell Lymphomas:

Diagnosis will be in line with Non-Hodgkin's Lymphoma: diagnosis and management NICE guideline NG52 <https://www.nice.org.uk/guidance/NG52/chapter/Recommendations#diagnosis>

Full body PET/CT scan is the recommended staging modality. CT neck/thorax/abdomen/pelvis with contrast is an alternative in exceptional/urgent clinical situations.

Diagnostic material (lymph node or soft tissue biopsies, bone marrow) is to be sent directly to Haematological Malignancy Diagnostic Service (HMDS) in Leeds. Lymph node excision biopsy is preferred, but a needle core biopsy using the largest calibre possible is an alternative.

The HMDS' T-lymphoid neoplasms reporting criteria are detailed in the link below (needs to log on to HMDS to access)

<https://www.hmids.leedsth.nhs.uk/docs/leeds/SOP/HMDS2184b%20supplementary%20to%20SOP%20R1-T-lymphoid%20neoplasms.pdf>

Patients with neurological symptoms and/or imaging studies suggestive of central nervous system involvement should undergo diagnostic lumbar puncture. CSF sample to be sent to HMDS, Leeds. Fertility-preserving treatments, such as sperm cryopreservation should be offered to eligible patients.

Management of T-Cell Lymphoma

T-Cell lymphoma will be managed within the Hull and North Lincolnshire MDT in line with Non-Hodgkin's Lymphoma: diagnosis and management NICE guideline NG52. <https://www.nice.org.uk/guidance/ng52>

The local management of peripheral T-Cell lymphomas will also take account of the following BSH and NICE guidelines:

The British Committee for Standards in Haematology (BCSH) Guidelines for the Management of Mature T-cell and NK-cell Neoplasms: Excluding cutaneous T-cell Lymphoma Updated August 2013

<https://b-s-h.org.uk/media/2895/t-nhl-guideline-3-8-13-updated-with-changes-accepted-v1-rg.pdf>

Management of Mature T-cell and and NK-cell Neoplasms published on the 11th of April 2011

<https://b-s-h.org.uk/guidelines/guidelines/management-of-mature-t-cell-and-and-nk-cell-neoplasms/>

Brentuximab vedotin for treating relapsed or refractory systemic anaplastic large cell lymphoma. Technology appraisal guidance [TA478] Published date: 04 October 2017

Brentuximab vedotin for treating CD30-positive cutaneous T-cell lymphoma. Technology appraisal guidance [TA577]Published date: 24 April 2019.

There is no consensus about optimal therapy for T-and NK-cell neoplasms, this is due to the rarity of these diseases and the lack of randomised trials. Therefore, current management decisions are based on small case series, phase II trials and expert opinion.

Patients should be entered into clinical trials wherever possible.

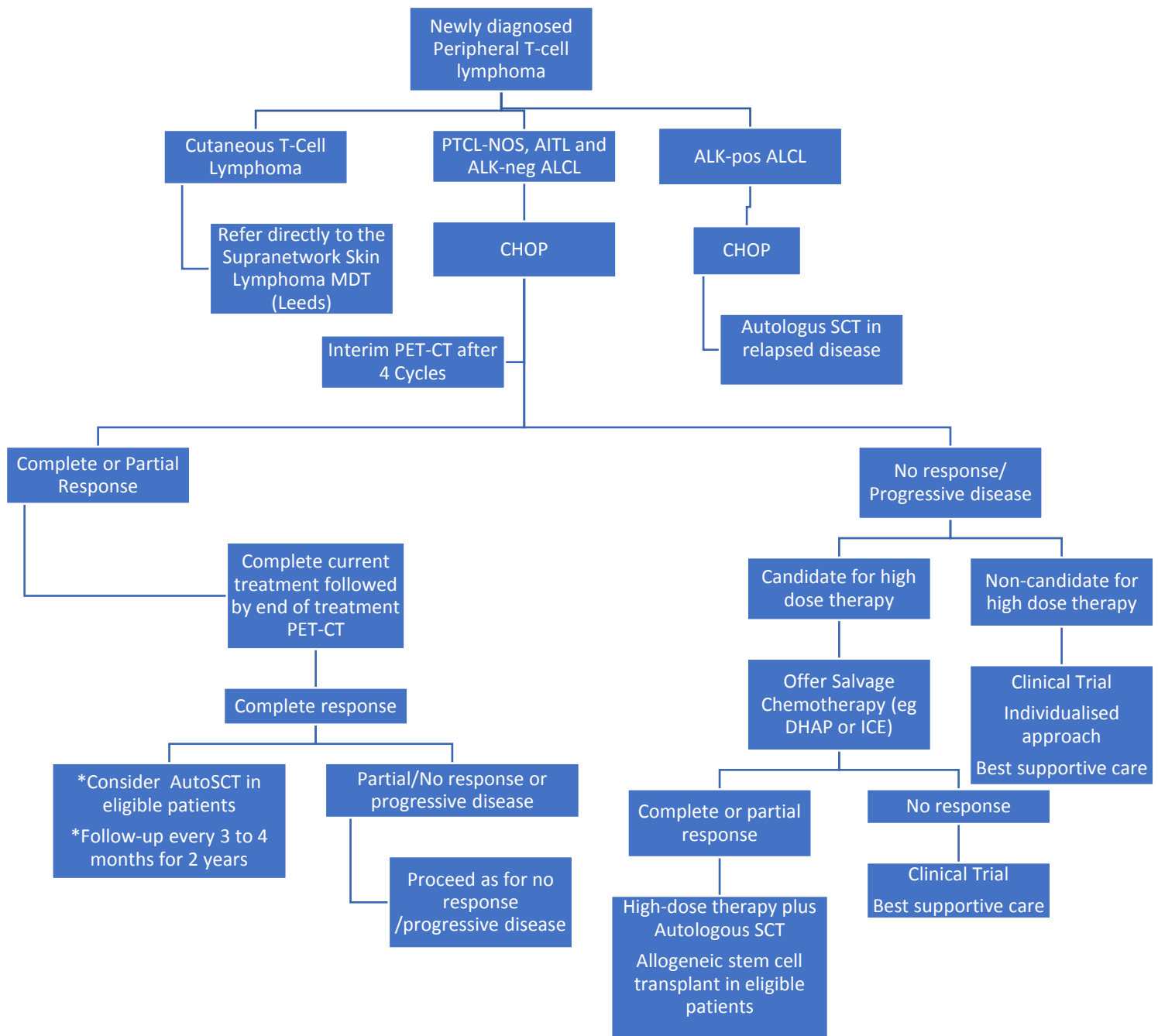
Peripheral T-Cell Lymphoma management algorithm

Newly Diagnosed Disease

- Not fit for intensive chemotherapy > best supportive care following MDT discussion
- Fit for intensive chemotherapy> CHOP
- Interim PET CT scan after 4 cycles
- Complete 6 cycles followed by end of treatment PET-CT and re-biopsy of any residual disease if possible.
- Complete response >>>>Consider consolidation with Autologous SCT in fit patients. Except for anaplastic lymphoma kinase (ALK)-positive anaplastic large cell lymphoma (ALCL) patients.
- ALK-positive ALCL is a chemosensitive malignancy and has outcomes comparable to, or better than, IPI adjusted DLBCL following anthracycline chemotherapy. Therefore ALK-pos ALCL should be treated in adults with CHOP chemotherapy as first line and platinum-based chemotherapy at relapse. Prognosis is so good in this group of patients that transplant should only be considered at relapse.
- Patients with limited stage anaplastic large cell lymphoma and no adverse prognostic features by IPI should be treated with 3-4 cycles of CHOP chemotherapy and involved field radiotherapy.
- Follow-up every 3 to 4 months for 2 years in those who achieve complete response.

Relapsed or refractory disease

- There is no data on which to base the choice of re-induction and the conventional approach is to use a platinum-based schedule (eg DHAP or ICE), particularly when intending to consolidate with a transplant.
- Offer salvage chemotherapy (DHAP (Dexamethasone / high-dose cytarabine/Cisplatin)/ ICE/ GDP/) to transplant eligible patients. Consolidate with Autologous SCT in those who achieve at least partial remission. Consider Allogeneic SCT in eligible patients.
- For transplant ineligible patients adopt an individualised approach via MDT discussion/ Palliative Treatment/ Best supportive care/ Clinical trial.
- Brentuximab Vedotin Regime: for Relapsed/refractory systemic anaplastic large cell lymphoma is NICE approved if: they have an Eastern Cooperative Oncology Group (ECOG) performance status of 0 or 1 and the company provides brentuximab vedotin according to the commercial access agreement with NHS England.
<https://www.nice.org.uk/guidance/ta478/chapter/1-Recommendations>
- Brentuximab Vedotin Regime for relapsed CD30 positive cutaneous T-cell lymphoma for IIB mycosis fungoides, primary cutaneous anaplastic large cell lymphoma or Sezary syndrome, in line with NICE guidance.
- This may be an option for some relapsed/refractory patients after MDT discussion
- A significant proportion of peripheral T-cell lymphomas (PTCL) express CD30 and may be potential candidates for CD30-targeting strategies but, this is yet to be approved.
- Central nervous system prophylaxis should be considered using the same criteria as for diffuse large B-cell NHL.



ALK= Anaplastic lymphoma kinase/ **PTCL-NOS**= Peripheral T-cell lymphoma, not otherwise specified/ **ALCL**= Anaplastic Large cell lymphoma/ **ICE**= Ifosfamide, Carboplatin, Etoposide/ **CHOP**= Cyclophosphamide, Doxorubicin, Vincristine, Prednisolone/ Angioimmunoblastic T-cell lymphoma (AITL)

References

- Haematology Multidisciplinary Team Operational Policy.
- Haematological cancers: improving outcomes NICE guideline [NG47]..
- Non-Hodgkin’s Lymphoma: diagnosis and management NICE guideline NG52.
- The British Committee for Standards in Haematology, Guidelines for the Management of Mature T-cell and NK-cell Neoplasms (Excluding cutaneous T-cell Lymphoma) Updated August 2013 .
- Brentuximab vedotin for treating relapsed or refractory systemic anaplastic large cell lymphoma Technology appraisal guidance [TA478] Published date: 04 October 2017.

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