

ONCOLOGY KNOWLEDGE HUB · FOR HEALTH PROFESSIONALS

T-cell Lymphomas

A mini review for clinicians

For postgraduate residents (medical, radiation and paediatric oncology, haematology, pathology, nuclear medicine, ENT, dermatology, medicine), practising clinicians and medical students

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Updated 17 October 2026. Educational summary written in our own words from current guidelines and primary trials; no drug doses; not advice for an individual patient. Verify current guidelines and Indian regulatory approvals before treatment decisions. Patients and families: please see the patient guides at drharitha.co.in.

T-cell Lymphomas

Haematological cancers, part 4 · PTCL-NOS, T follicular helper (angioimmunoblastic), systemic anaplastic large cell (ALK+/ALK-), extranodal NK/T, ATLL and T-PLL · CD30 and BV-CHP · Consolidation · Relapse · Key points for clinicians · Last updated 17 October 2026

01 At a glance

- **Peripheral T-cell lymphomas (PTCL) are rare and varied:** about 10% of non-Hodgkin lymphomas in the West and 15–20% in Asia. The main systemic types are PTCL, NOS; nodal T follicular helper (TFH) lymphoma, angioimmunoblastic type (AITL); systemic anaplastic large cell lymphoma (ALCL, ALK-positive or ALK-negative); extranodal NK/T-cell lymphoma, nasal type; ATLL; and T-PLL.
- **Diagnosis needs an excision biopsy and an expert haematopathologist:** PTCL-NOS is a diagnosis of exclusion; CD30, ALK, TFH markers, EBER and HTLV-1 testing name the entity.
- **CD30 chooses first-line treatment:** CD30-positive (at least 1% of cells, including all ALCL) → brentuximab vedotin with CHP ([ECHELON-2^{\[1\]}](#)); CD30-negative → CHOEP if fit and about 65 or younger, CHOP otherwise.
- **Outcome depends on the entity:** ALK-positive ALCL does well (3-year OS about 90%); PTCL-NOS, AITL and ALK-negative ALCL relapse often; relapsed PTCL without transplant has a median survival of about 6 months.
- **Response-guided care:** PET after three cycles and at the end; consolidation (RT for limited residual disease; autologous transplant for selected fit patients) is decided at the end of induction.
- **Relapse:** biopsy; BV for BV-naïve ALCL, crizotinib for ALK-positive ALCL, single agents (belinostat, pralatrexate, romidepsin) or combination salvage for PTCL, and transplant as the only route to long-term control.
- **India:** ALCL is relatively frequent (4.3% of NHL at Tata Memorial), and PTCL and nasal NK/T lymphoma are more common than in the West: a necrotic nasal mass needs a deep biopsy.

Peripheral T-cell lymphomas: rare, varied, and harder to cure than B-cell lymphomas



Widths illustrative. PTCLs are about 10% of NHL in the West and 15–20% in Asia; skin-based T-cell lymphomas are covered in their own module.

What they are

- Mature (post-thymic) T- or NK-cell cancers
- Mostly older adults; B symptoms common
- Diagnosis needs an excision biopsy and an expert pathologist

How they are treated

- CD30+ (≥1% of cells): BV-CHP; CD30-: CHOEP if fit, CHOP if not
- Interim and end-of-treatment PET
- Selected patients: autologous transplant in first remission

What to expect

- ALK+ ALCL: 3-year OS about 90%
- Most other PTCLs: relapse is common
- Relapse: BV, ALK inhibitor, single agents, transplant

Figure 1. T-cell lymphomas at a glance (UpToDate PTCL-NOS, AITL, sALCL, ENKTL, ATLL and T-PLL reviews).

02 Clinical presentation, common traps & causes

Six systemic T- and NK-cell lymphomas and the clue to each

PTCL, NOS Diagnosis of exclusion; nodes ± extranodal; B symptoms, eosinophilia, itching, HLH	TFH / AITL Sudden illness: nodes, liver, spleen, fever, itchy rash, arthritis, high globulins, haemolysis	ALCL, ALK+ Children and young adults; fast-growing nodes, skin, bone; NPM::ALK	ALCL, ALK- Older adults; same look, worse outlook; DUSP22 rearranged in up to a third
NK/T, nasal type Destructive nose, sinus or palate mass; necrosis; EBV in every case	ATLL HTLV-1; flower cells, high calcium, skin lesions; Caribbean, Japan, Peru, Iran	T-PLL Older adult, very high count (often >100,000/μL), spleen, skin, effusions	Not here Mycosis fungoides, Sézary, cutaneous ALCL, SPTCL: cutaneous module

India: anaplastic large cell lymphoma was 4.3% and other nodal PTCL 2.9% of NHL at Tata Memorial; PTCL and nasal NK/T lymphoma are less common than in East Asia but more common than in the West. A nasal ulcer or "non-healing sinusitis" with necrosis needs a deep biopsy, not more antibiotics.

Figure 2. Entities and clues (UpToDate diagnosis reviews); Indian data from Naresh 2000 and a 2020 Bihar series.

Who gets it, and why

- **PTCL-NOS**: median age about 60; the commonest PTCL outside Asia (about a third of cases); in Asia, NK/T-cell lymphoma is commoner.
- **AITL**: older adults with an acute systemic illness: generalised nodes (76–95%), splenomegaly (70%), B symptoms (70–85%), itchy rash (20–60%), arthritis, effusions; 90% stage III–IV; polyclonal hypergammaglobulinaemia, autoimmune haemolysis.
- **ALCL**: about 2% of adult NHL; ALK-positive disease in children and young adults, ALK-negative in older adults; male predominance; fast-growing nodes with skin, bone, liver, lung or CNS disease.
- **NK/T-cell lymphoma**: EBV in every case; commonest in East Asia and Central and South America; nose, sinus and palate (stage I 43%, II 22% in one series).
- **ATLL**: HTLV-1 carriers, after decades of latency (lifetime risk 4–5% in endemic areas: southwest Japan, the Caribbean, west Africa, Peru, northeast Iran). **T-PLL**: older adults; much more common with ataxia-telangiectasia.
- **India**: in 2773 lymphomas at Tata Memorial, ALCL was 4.3% and other nodal PTCL 2.9% of NHL; ALCL and T-lymphoblastic lymphoma were relatively commoner than in the West ([Naresh 2000^{\[2\]}](#)); a Bihar series found PTCL and nasal NK/T lymphoma less common than in some Asian countries but commoner than in the West ([Bihar 2020^{\[3\]}](#)).

Common traps

- **Fever, nodes and rash treated as infection or drug allergy for weeks.** AITL looks like a systemic infection or autoimmune disease: biopsy a node early.
- **"Reactive" or "atypical lymphoid infiltrate" on a core.** T-cell lymphomas need architecture and a large panel: excise a whole node.
- **A necrotic nasal ulcer called fungal sinusitis or granulomatosis.** Superficial biopsies show only necrosis; ask for deep biopsies and EBER.

- **CD30 not reported.** It decides first-line treatment: ask for it on every PTCL.
- **Hypercalcaemia and "Sézary-like" cells in a patient from an HTLV-1 area.** Test HTLV-1 before calling it mycosis fungoides or PTCL.
- **Missing HLH.** Fever, cytopenias, very high ferritin: hemophagocytic lymphohistiocytosis can complicate PTCL and needs urgent treatment.

SHARE WITH PATIENTS: SYMPTOMS, SWELLINGS AND DIAGNOSIS

Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

Every swollen lymph node is cancer?

If there is no pain, fever or weight loss, lymphoma is impossible?

Itching all over the body is always just a skin problem?

FNAC alone can reliably diagnose every lymphoma?

Lymphoma can't occur outside the lymph nodes?

All lymphomas are the same cancer?

03 Diagnosis, workup & staging essentials

Making the diagnosis

- **Excision or incision biopsy** of a node (or deep biopsies of the involved organ); fresh tissue for flow cytometry where possible; expert haematopathology review.
- **PTCL-NOS**: one or more pan-T antigens (CD2, CD3, CD5, CD7), often with loss of CD5 or CD7; clonal TCR; no specific cytogenetics; diagnosis only after excluding other types.
- **TFH lymphoma (AITL)**: effaced node, arborising high endothelial venules, expanded CD21-positive dendritic meshworks; CD4 plus at least two TFH markers (PD-1, ICOS, CXCL13, BCL6, CD10); EBV-positive B blasts are common.
- **ALCL**: sheets of large "hallmark" cells with strong membrane and Golgi CD30; ALK by IHC or FISH (NPM::ALK) separates ALK-positive from ALK-negative; DUSP22 rearrangement in up to a third of ALK-negative cases. Exclude primary cutaneous and breast implant-associated ALCL.
- **NK/T-cell lymphoma**: necrosis, angioinvasion, CD56, cytoplasmic CD3, cytotoxic granules, and EBER in virtually all.
- **ATLL**: flower cells, CD4+CD25+CD7-, and HTLV-1 serology (confirmatory blot or PCR); at least 5% abnormal T cells in blood plus HTLV-1 can establish leukaemic forms ([Shimoyama 1991](#)^[4], [Cook 2019](#)^[5]).
- **T-PLL**: over 5000/μL clonal T cells with TCL1 or MTCPI abnormality (inv(14), t(14;14), t(X;14)); HTLV-1 negative ([Staber 2019](#)^[6]).

The T-cell lymphoma key: markers that name the entity

	Defining markers	Genetics / virus
PTCL, NOS	One or more pan-T antigens (CD2, CD3, CD5, CD7); often loses CD5 or CD7	No specific change; clonal TCR; diagnosis of exclusion
TFH / AITL	CD4 + two of PD-1, ICOS, CXCL13, BCL6, CD10; CD21 dendritic meshwork	TCR clonal in 75–90%; EBV+ B blasts; IgH clonal in 25%
ALCL	CD30 strong, membrane + Golgi; T antigens variable; no B markers	ALK by IHC or FISH (NPM::ALK); ALK-: DUSP22 in up to a third
NK/T, nasal	CD2, CD56, cytoplasmic CD3, granzyme/TIA-1; surface CD3 negative	EBER positive in virtually all; TCR usually germline
ATLL	CD4, CD25 (most), CD7 lost; dim CD3; FoxP3 in some	HTLV-1 antibodies or provirus PCR
T-PLL	CD7, CD52 strong; CD4+CD8+ in a quarter; TCL1 overexpressed	inv(14)/t(14;14)/t(X;14); HTLV-1 negative

Every case: excision biopsy, expert review, CD30 (it chooses first-line treatment), and HTLV-1 serology where relevant. EBER separates NK/T lymphoma.

Figure 3. T-cell lymphoma key (UpToDate PTCL-NOS, AITL, sALCL, ENKTL, ATLL and T-PLL diagnosis reviews; simplified).

Before treatment

Area	What to do	Why
History and examination	B symptoms, rash, nasal and palatal examination, skin, liver, spleen, performance status, comorbidities, country of origin (HTLV-1)	Entity, stage, fitness

Area	What to do	Why
Blood tests	CBC and smear, chemistry, calcium, LDH, uric acid, immunoglobulins, DAT, ferritin if febrile and cytopenic	IPI/PIT; hypercalcaemia (ATLL); haemolysis (AITL); HLH
Viruses	HBV, HCV, HIV; HTLV-1 where relevant; EBV (EBER on tissue)	Prophylaxis; ATLL; NK/T diagnosis
Imaging	PET-CT; MRI of nose and sinuses for NK/T lymphoma	Stage; baseline for interim and end PET
Marrow	Biopsy when it changes management (cytopenias, PIT, T-PLL)	PIT score; stage
Heart and fertility	Echocardiogram before anthracyclines; sperm banking, fertility counselling	CHOP-based treatment; transplant plans
Transplant assessment	Fitness and donor search planned early for fit patients	Relapses come early

Prognosis: ALK status matters most; the scores help

3-year overall survival in systemic ALCL (%)



PIT: one point each

- Age over 60
- LDH above normal
- ECOG 2 or more
- Bone marrow involvement

IPI (age, LDH, ECOG, stage, extranodal sites) also works; in one PTCL series 45% scored 3 or more. Stage for stage, PTCL does worse than B-cell lymphoma. AITL has its own score; ENKTL, ATLL and T-PLL have disease-specific indices. ALK-positive disease is mostly young patients: age explains part of the gap.

Relapsed PTCL not going to transplant: median overall survival about 6 months in one series. Hence: trials, and consolidation for selected patients.

Figure 4. Prognosis (Gallamini 2004, PIT; Vose 2008, International T-cell Project; UpToDate PTCL-NOS and sALCL reviews).

SHARE WITH PATIENTS: SCANS, BIOPSIES AND STAGE

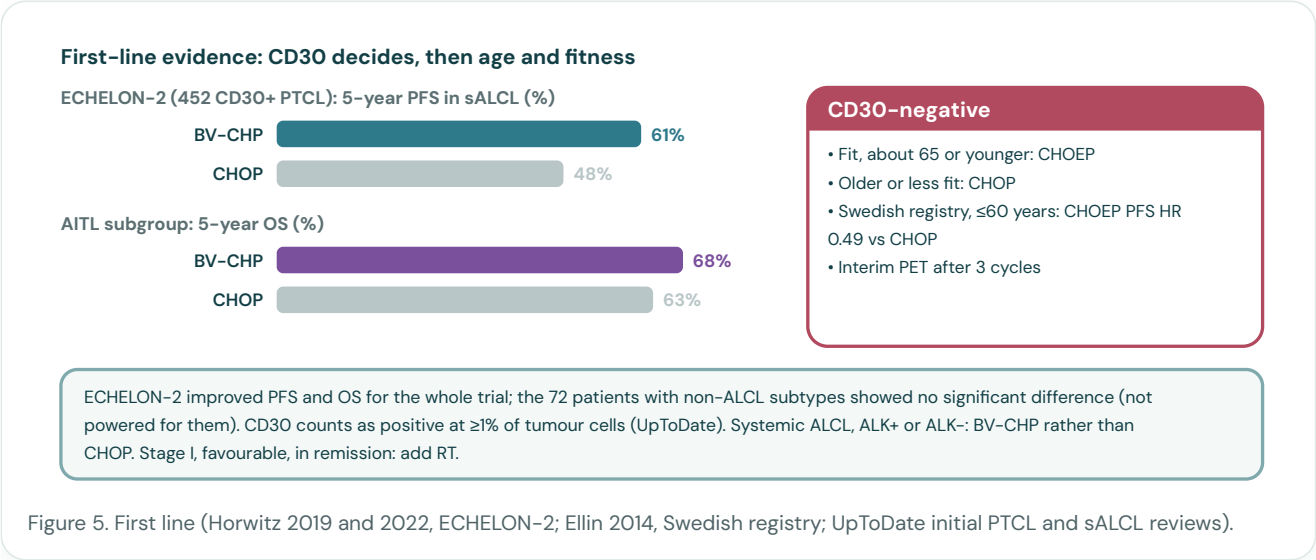
Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

[A lymph node that lights up on PET always means cancer?](#)

[Stage IV lymphoma means the disease is incurable?](#)

o4 Biology and biomarkers that change treatment

Marker or feature	What it tells you	What it changes
CD30 (≥1% of tumour cells)	All ALCL; a share of PTCL-NOS and AITL	BV-CHP first line; BV at relapse
ALK (IHC or FISH)	ALK-positive ALCL: young, curable (3-year OS about 90% vs 62%)	No transplant in first CR; crizotinib at relapse
DUSP22 rearrangement (ALK-ALCL)	Up to a third; prognostic meaning debated	Not used for treatment choice yet
TFH markers (PD-1, ICOS, CXCL13, BCL6, CD10)	Nodal TFH lymphoma (AITL and related)	Same first-line approach; azacitidine studied at relapse (ORACLE^[7])
EBV (EBER)	NK/T-cell lymphoma (all); EBV+ B blasts in AITL	NK/T diagnosis; watch for EBV-driven B-cell proliferations
HTLV-1	ATLL in every case	Different disease, different treatment; family screening
TCL1 / MTCPI, inv(14)	T-PLL	Distinguishes it from ATLL and Sézary



05 Local therapy: radiotherapy & surgery

Radiotherapy

- **Stage I PTCL with a favourable prognosis** in complete or partial response after chemotherapy: adjuvant RT.
- **Limited-stage ALCL** in complete response: observation or RT; limited residual disease: RT to convert the response, then transplant in eligible ALK-negative patients.
- **Advanced disease with a single residual site**: RT, followed by autologous transplant in fit patients with PTCL-NOS, AITL or ALK-negative ALCL; RT alone for ALK-positive ALCL and for patients who cannot have a transplant.
- **NK/T-cell lymphoma**: RT is part of combined-modality treatment for localised disease (about 50 Gy ISRT in sandwich protocols; inadequate RT dose is a cause of local failure); RT alone for less-fit patients with localised disease.

Surgery

- **Diagnosis**: excision biopsy is the main role; deep biopsies for nasal disease.
- **Emergencies**: perforation or bleeding from intestinal T-cell lymphoma; airway compromise from nasal or pharyngeal disease.

o6 Systemic therapy: indications & options

PTCL-NOS, TFH lymphoma (AITL) and EATL: first line

- **CD30-positive (≥1%):** BV-CHP for six cycles. In ECHELON-2 (452 patients, mostly ALCL), BV-CHP improved PFS and OS over CHOP; in the 72 patients with other subtypes the gain was not significant (AITL 5-year OS 68% vs 63%) ([ECHELON-2^{\[1\]}](#), [five-year update^{\[8\]}](#)).
- **CD30-negative, fit, about 65 or younger:** CHOEP; in the Swedish registry CHOEP gave better PFS than CHOP under 60 (HR 0.49) ([Ellin 2014^{\[9\]}](#), [Schmitz 2010^{\[10\]}](#)). **Older or less fit:** CHOP.
- **Interim PET after three cycles:** CR or PR → complete six cycles; stable or progressive → treat as refractory.
- **After induction** (end-of-treatment PET): stage I favourable → RT; advanced, fit, in CR → observation or autologous transplant ([NLG-T-01^{\[11\]}](#)); single residual site → RT then autologous transplant; extensive residual → refractory pathway. Not transplant-eligible → RT for CR or limited residual disease.

After induction: who needs a transplant in first remission?

ALK+ ALCL, CR	ALK- ALCL, PTCL-NOS, AITL: fit	Not transplant-eligible
• Observe • Limited residual disease: RT • Transplant only at relapse	• CR: observe or autologous HCT • Limited residual: RT then auto-HCT • Extensive residual: treat as refractory	• CR or limited residual: RT • More: treat as refractory • Stage I favourable: RT after chemo

Auto-HCT in first remission: overall survival in prospective vs retrospective studies (%)



No randomised trial proves a survival gain from transplant in first remission; selection bias inflates retrospective results. Allogeneic HCT is kept for relapse.

Figure 6. Consolidation (d'Amore 2012, NLG-T-01; meta-analyses as quoted in the UpToDate initial PTCL and sALCL reviews).

Systemic anaplastic large cell lymphoma: first line

- **BV-CHP for every systemic ALCL** (ALK-positive or negative) rather than CHOP: in the 316 ALCL patients of ECHELON-2, 5-year PFS 61% vs 48% (HR 0.55) with a trend to better survival.
- **Interim PET after 3–4 cycles**, then complete six cycles if responding.
- **ALK-positive, advanced, CR:** observe (no transplant); limited PR → RT. **ALK-negative, advanced, CR:** observation or autologous transplant; limited PR → RT then autologous transplant. Extensive PR → relapsed/refractory pathway.
- **Frail patients:** single-agent BV. **Children:** clinical trials or paediatric protocols.

Relapsed or refractory PTCL and ALCL

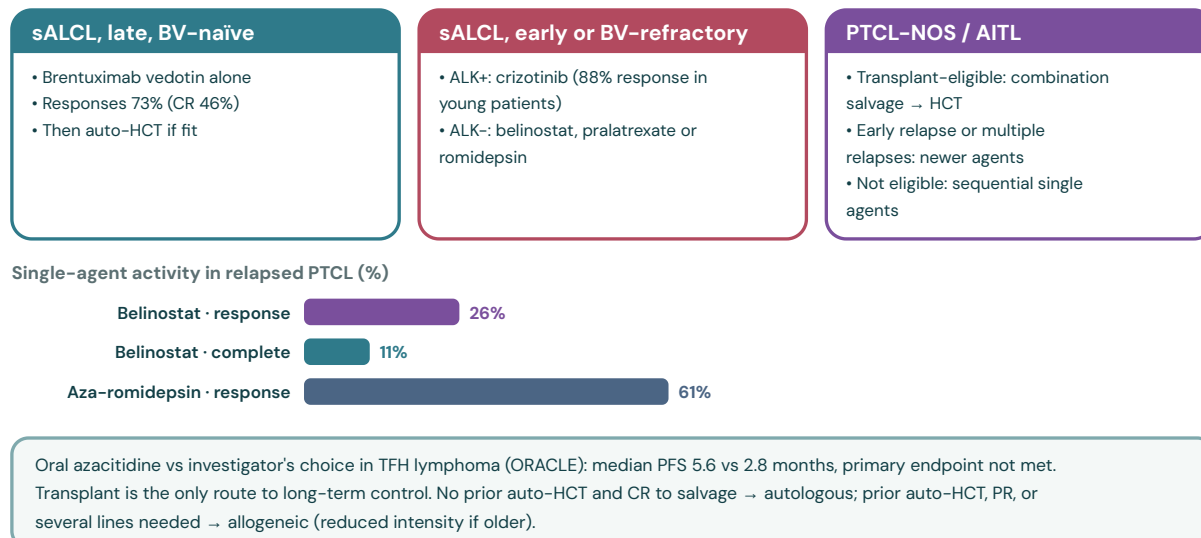
Relapsed or refractory disease: biopsy, then choose by ALK, BV exposure and transplant plan

Figure 7. Relapsed disease (Pro 2012 and 2017, BV; Mossé 2013 and paediatric crizotinib series; O'Connor 2015, BELIEF; Falchi 2021; Dupuis 2024, ORACLE; UpToDate reviews).

- **Biopsy** to confirm relapse and check for a change in histology; trials whenever possible.
- **sALCL, relapse after 6 months or BV-naïve refractory:** BV alone (73% response, 46% complete in BV-naïve patients) ([Pro 2012](#)^[12], [Pro 2017](#)^[13]). **Early or BV-refractory:** ALK-positive → crizotinib (88% response in patients aged 21 or under; [Mossé 2013](#)^[14]); ALK-negative → belinostat, romidepsin or pralatrexate.
- **After response:** ALK-positive, no prior transplant → autologous transplant; prior autologous transplant → observe. ALK-negative → autologous, allogeneic or observation.
- **PTCL-NOS and AITL, transplant-eligible:** combination salvage as a bridge; CR without prior autologous transplant → autologous; prior autologous transplant, PR, or several lines needed → allogeneic (reduced intensity if older).
- **Primary refractory, relapse within 6 months, or multiple relapses:** a newer agent rather than combination chemotherapy. **Not transplant-eligible:** sequential single agents (belinostat 26% response; [BELIEF](#)^[15], [PROPEL](#)^[16]). Oral azacitidine for TFH lymphoma did not meet its endpoint (median PFS 5.6 vs 2.8 months; [ORACLE](#)^[7]); azacitidine-romidepsin was active in a phase 2 study ([Falchi 2021](#)^[17]).

Extranodal NK/T-cell lymphoma, nasal type

Extranodal NK/T-cell lymphoma, nasal type**Presentation**

- Nasal blockage, bleeding, destructive mass of nose, sinus, palate
- Stage I 43%, II 22%; B symptoms 37%
- Also skin, gut, testis, airway

Pathology

- Necrosis and angioinvasion: take deep, large biopsies
- CD56, cytoplasmic CD3, cytotoxic granules
- EBER positive: required

Mimics

- Fungal or TB granuloma, granulomatosis with polyangiitis
- Squamous and nasopharyngeal carcinoma
- Superficial biopsies show only necrosis

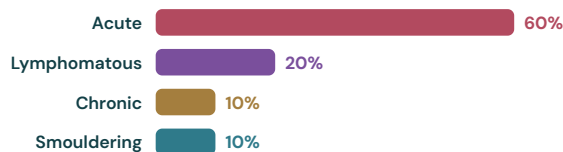
Localised (IE, contiguous IIE): combined chemotherapy (asparaginase- or platinum-based) and RT, concurrent or sandwich; CR → observe; less than CR → transplant. Advanced or non-nasal: asparaginase-based induction (m-SMILE, DDGP, P-GEMOX) → transplant in CR or PR. Relapse: PD-1 antibody or chemotherapy, then transplant.

Not anthracyclines: CHOP-like regimens do poorly. Follow with endoscopy, PET-CT and plasma EBV DNA; scores: PINK, PINK-E.

Figure 8. ENKTL (UpToDate ENKTL diagnosis and treatment reviews; Kim 2016, PINK; Yamaguchi 2009 and 2011; Kim 2009; Kwong 2017).

- **Evaluate** at an experienced centre: examination, fibre-optic endoscopy, PET-CT, marrow, plasma EBV DNA. Localised = stage IE or contiguous IIE (cervical nodes); advanced = non-contiguous II, III–IV and all non-nasal disease. Prognosis: PINK and PINK-E (with EBV load) ([Kim 2016](#)^[18]).
- **Localised, fit**: combined-modality treatment with asparaginase- or platinum-based chemotherapy and RT rather than anthracyclines, chemotherapy alone or RT alone. Options: concurrent chemoradiotherapy with reduced-dose DeVIC ([JCOG0211](#)^[19]) or RT with cisplatin followed by VIDL/VIPD ([Kim 2009](#)^[20]); or sequential/sandwich treatment with modified SMILE or P-GEMOX around ISRT of about 50 Gy. Systemic failure still occurs in 25–50%.
- **After treatment**: CR → observation; less than CR → transplant (the only reliable cure in this setting), with bridging if needed.
- **Advanced**: asparaginase-based induction (modified SMILE, DDGP or P-GEMOX; [SMILE](#)^[21]), then transplant for CR or PR.
- **Less fit**: localised → RT alone or lower-intensity CMT (GDP or MGAD); advanced → GDP, with RT for bulky symptomatic disease.
- **Relapsed or refractory**: salvage before transplant; a PD-1 antibody before planned autologous transplant (pembrolizumab highly active: [Kwong 2017](#)^[22]), asparaginase or other chemotherapy otherwise; transplant is the only proven cure.

ATLL and T-PLL

Leukaemic T-cell neoplasms: ATLL and T-PLL**ATLL clinical variants (% of cases)****T-PLL criteria (all 3 major, or 2 + 1 minor)**

- Over 5000/μL T-PLL-phenotype cells in blood or marrow
- Clonal T cells (TCR PCR or flow)
- 14q32 or Xq28 abnormality, or TCL1/MTCP1 expression
- Minor: chromosome 11 (ATM), 8, 5, 12, 13, 22; spleen or effusions

ATLL: HTLV-1 test in every suspected case; hypercalcaemia (up to 70% in acute), lytic bone, opportunistic infections incl. Strongyloides; acute and lymphomatous: survival in months. ATLL: acute, lymphomatous, unfavourable chronic → VCAP-AMP-VECP (± mogamulizumab) or CHOP if older, intrathecal prophylaxis, allogeneic HCT if a donor; chronic, smouldering → watch. T-PLL: inactive (15–20%) → watch with monthly counts; active → IV alemtuzumab (CR in most), add pentostatin if suboptimal, then HCT in CR (allogeneic if young and fit).

Figure 9. ATLL and T-PLL (UpToDate ATLL and T-PLL diagnosis and treatment reviews; Staber 2019; Tsukasaki 2007; Dearden 2001).

- **ATLL, acute, lymphomatous or unfavourable chronic:** VCAP-AMP-VECP (better than biweekly CHOP; [JCOG9801](#)^[23]), with mogamulizumab where available (CR 52% vs 33%, median PFS 8.5 vs 6.3 months; [Ishida 2015](#)^[24]); avoid mogamulizumab before allogeneic transplant (graft-versus-host risk). Older or comorbid: CHOP or CHOP-like. Always intrathecal prophylaxis. Trials encouraged.
- **After remission:** allogeneic transplant for patients with a donor (graft-versus-leukaemia effect). Zidovudine plus interferon alfa has been used, with median survival of 6–18 months in reported series ([Bazarbachi 2010](#)^[25]).
- **Typical chronic and smouldering ATLL:** no immediate treatment; median survival of smouldering disease untreated is about 3 years. Treat infections and hypercalcaemia; prophylaxis against opportunistic infections.
- **T-PLL, inactive:** close observation with monthly blood counts. **Active** (B symptoms, enlarging nodes, spleen or liver, marrow failure, extramedullary disease, rapidly rising count): IV alemtuzumab alone; add pentostatin if progression, stable disease at 4–6 weeks or less than CR at 10–12 weeks ([Dearden 2001](#)^[26]).
- **T-PLL consolidation:** transplant in CR (allogeneic for younger fit patients; autologous if not a candidate). Relapse: trials. Median survival with modern treatment is 1–3 years.

India now (October 2026)

- Indian frequencies (Tata Memorial 2000; Bihar 2020) are web-verified; HTLV-1 is not endemic in India, but ATLL is seen in people from endemic regions.
- Availability and cost of brentuximab vedotin, crizotinib, belinostat, pralatrexate, romidepsin, pegaspargase, PD-1 antibodies, mogamulizumab and alemtuzumab in India, and access to transplant, to be confirmed by the treating team.

SHARE WITH PATIENTS: TREATMENT AND RELAPSE

Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

If lymphoma returns after chemotherapy, no effective treatment remains?

Radiotherapy has no role in lymphoma?

Immunotherapy and targeted therapy are the same as chemotherapy?

Aggressive lymphoma is always harder to cure than slow-growing lymphoma?

Quick reference (also as a printable PDF on the Printable Teaching Resources page)

Table A. Diagnosis and staging tests and what they change

Test	When	What it changes	Pitfalls
Excision biopsy (deep biopsies for nasal disease)	Every suspected case	Entity	Cores and superficial biopsies miss architecture or show only necrosis
IHC: CD3, CD4, CD8, CD30, ALK, PD-1/ICOS/CXCL13, CD56, granzyme; EBER	Every biopsy	ALCL ALK±, TFH lymphoma, NK/T, PTCL-NOS	CD30 must be quantified (≥1% counts)
HTLV-1 serology (blot or PCR)	Abnormal T cells in blood, hypercalcaemia, origin in endemic areas	ATLL	Healthy carriers can have flower cells
Flow cytometry, TCL1, karyotype/FISH	High T-cell counts	T-PLL vs ATLL vs Sézary	TCL1-negative T-PLL exists (minor criteria)
PET-CT; MRI nose and sinuses	Staging; interim (after 3 cycles) and end of treatment	Stage; response-guided plan	Inflammation and infection take up FDG
LDH, calcium, ferritin, DAT, immunoglobulins	Baseline	IPI/PIT; ATLL; HLH; haemolysis in AITL	Ferritin very high with cytopenias: think HLH
HBV, HCV, HIV	Before treatment	Prophylaxis	—

Table B. Treatment by setting

Setting	Standard choice	Key evidence	Notes
CD30-positive PTCL (incl. all systemic ALCL)	BV-CHP × 6	ECHELON-2^[1]	Benefit clearest in ALCL
CD30-negative PTCL, fit ≤65	CHOEP × 6	Ellin 2014^[9]	Interim PET after 3 cycles
CD30-negative PTCL, older or less fit	CHOP × 6	UpToDate initial PTCL review	RT for residual disease
PTCL/ALK- ALCL, advanced, fit, CR	Observation or autologous HCT	NLG-T-01^[11]	No randomised proof of survival gain
ALK+ ALCL, CR	Observation	UpToDate sALCL review	No transplant in first CR
Stage I favourable PTCL, CR/PR	Adjuvant RT	UpToDate initial PTCL review	—

Setting	Standard choice	Key evidence	Notes
Relapsed sALCL, late or BV-naïve	BV	Pro 2012 ^[12]	Then HCT if fit
Relapsed ALK+ ALCL, early or BV-refractory	Crizotinib	Mossé 2013 ^[14]	Young patients best studied
Relapsed PTCL, transplant-eligible	Combination salvage → auto or allo HCT	UpToDate relapsed PTCL review	Newer agent if early or multiple relapse
Relapsed PTCL, not eligible	Sequential single agents	BELIEF ^[15] , PROPEL ^[16]	Trials; palliative care
NK/T, localised	Asparaginase- or platinum-based chemotherapy + RT (concurrent or sandwich)	JCOG0211 ^[19] , Kim 2009 ^[20]	No anthracyclines; CR → observe
NK/T, advanced	m-SMILE, DDGP or P-GEMOX → HCT	SMILE ^[21]	Relapse: PD-1 antibody before auto-HCT
ATLL, aggressive types	VCAP-AMP-VECP ± mogamulizumab (or CHOP if unfit), IT prophylaxis → allo-HCT	JCOG9801 ^[23] , Ishida 2015 ^[24]	No mogamulizumab before allo-HCT; chronic/smouldering: watch
T-PLL, active	IV alemtuzumab (+ pentostatin if suboptimal) → HCT in CR	Dearden 2001 ^[26]	Inactive: watch with monthly counts

Table C. Regimens and drugs: evidence and best fit

Regimen or drug	Best fit	Evidence	Main toxicity
BV-CHP	CD30-positive PTCL, all systemic ALCL	ECHELON-2: sALCL 5-year PFS 61% vs 48%	Neuropathy, neutropenia
CHOEP	Fit, ≤65, CD30-negative	Swedish registry: PFS HR 0.49 vs CHOP (≤60)	More myelosuppression than CHOP
CHOP	Older or less fit	Historical standard	Neutropenia, neuropathy, cardiotoxicity
Brentuximab vedotin alone	Relapsed CD30+ disease; frail sALCL	Relapsed sALCL: 73% response, 46% complete	Peripheral neuropathy
Crizotinib	Relapsed ALK+ ALCL	88% response in patients ≤21 years	See product information
Belinostat / pralatrexate / romidepsin	Relapsed PTCL, ALK-ALCL	Belinostat: 26% response (11% complete)	Belinostat: nausea (42%), fatigue, fever; cytopenias
Oral azacitidine (± romidepsin)	Relapsed TFH lymphoma	ORACLE: PFS 5.6 vs 2.8 months (not significant); combination 61% response (phase 2)	Cytopenias, GI effects
Autologous / allogeneic HCT	Responding relapse; selected first CR	Only route to long-term control after relapse	Transplant-related mortality (allo)

Table D. Managing problems during treatment

Problem	Prevent	Recognise	Act
Hypercalcaemia (ATLL)	Check calcium at diagnosis and during treatment	Confusion, thirst, constipation, kidney injury	IV fluids and calcium-lowering treatment; treat the lymphoma
HLH	High suspicion in febrile cytopenic patients	Fever, splenomegaly, ferritin very high, falling fibrinogen	Urgent specialist treatment alongside the lymphoma
Peripheral neuropathy (BV, vincristine)	Ask about tingling every cycle	Numbness, weakness, falls	Dose delay or reduction per protocol
Hepatitis B reactivation	HBsAg and anti-HBc before treatment	Rising ALT, HBV DNA	Antiviral prophylaxis; hepatology
Infections (incl. Strongyloides in ATLL)	Screen and treat Strongyloides before steroids in people from endemic areas; non-live vaccines	Fever, diarrhoea, chest signs	Early antibiotics; specialist advice
Tumour lysis	Hydration ± allopurinol in bulky disease	Rising potassium, phosphate, uric acid, creatinine	Monitor closely in the first days

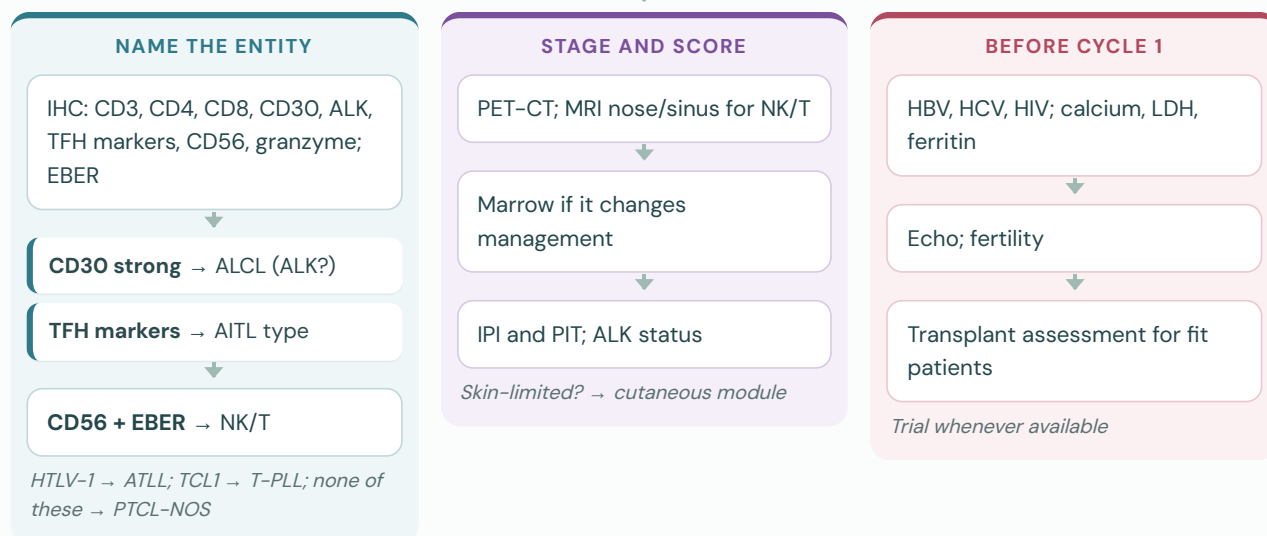
07 Management algorithms

Five decision paths pull together sections 02–06.

ALGORITHM 1

Suspected T-cell lymphoma: to an entity and a plan

Nodes with fever, rash or itching; a necrotic nasal mass; or abnormal T cells in blood → excision or deep biopsy before steroids; HLH screen if febrile and cytopenic



All: expert haematopathology · CD30 reported on every case · plan the end-of-treatment decision at the start.

ALGORITHM 2

PTCL-NOS, AITL and EATL: first line

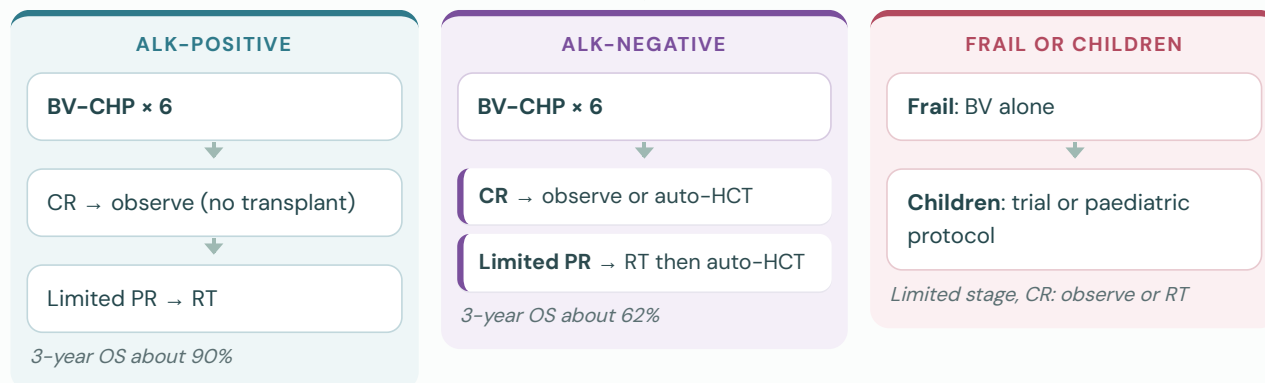


End of treatment: stage I favourable → RT · fit, CR → observe or auto-HCT · single residual site → RT then auto-HCT · extensive residual → refractory pathway.

ALGORITHM 3

Systemic ALCL by ALK status

Systemic ALCL (exclude cutaneous and breast implant-associated) → ALK, stage, fitness

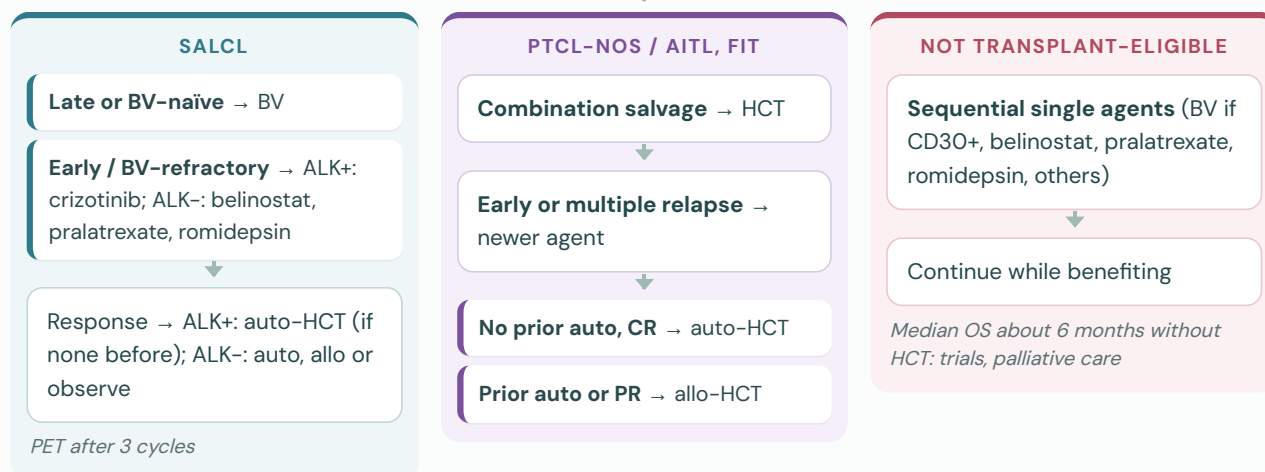


All: interim PET after 3–4 cycles · extensive PR or progression → relapsed/refractory pathway.

ALGORITHM 4

Relapsed or refractory PTCL and ALCL

Suspected relapse → biopsy (histology may change) → PET → transplant plan



All: clinical trial first · infection prophylaxis and vaccines · early palliative care alongside treatment.

ALGORITHM 5

Extranodal NK/T-cell lymphoma

NK/T-cell lymphoma confirmed (EBER+) → endoscopy, PET-CT, marrow, plasma EBV DNA → stage and fitness

LOCALISED (IE / CONTIGUOUS IIE)

CMT: concurrent CRT (DeVIC or cisplatin → VIDL) or sandwich (m-SMILE, P-GEMOX + RT)

CR → observe

Less than CR → HCT

Less fit: RT alone or GDP/MGAD + RT

ADVANCED OR NON-NASAL

Asparaginase-based: m-SMILE, DDGP or P-GEMOX

CR or PR → HCT

Less fit: GDP ± RT to bulky sites

RELAPSED OR REFRACTORY

Salvage before transplant

Auto-HCT planned → PD-1 antibody

Allo-HCT planned → asparaginase-based if not recent

Transplant is the only proven cure

All: no anthracycline-based regimens · response by endoscopy, PET-CT and EBV DNA · experienced centre and trials.

o8 Survivorship, follow-up & late effects

A simple follow-up rhythm after treatment of T-cell lymphoma

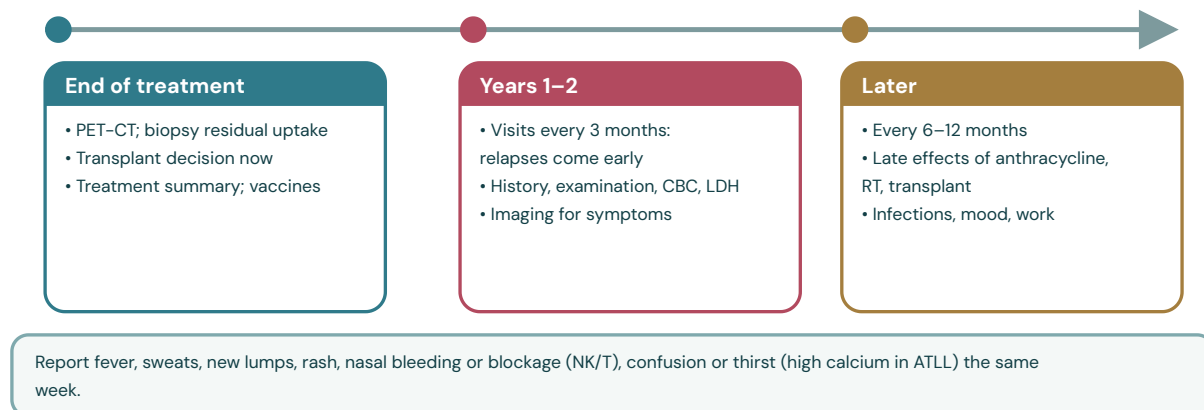


Figure 10. A simple follow-up rhythm, adapted from the UpToDate PTCL and sALCL treatment reviews. Individualise.

- **End of treatment:** PET-CT; biopsy residual uptake if it would change management; record the transplant decision.
- **Years 1–2:** relapses come early: visits every 3 months with history, examination, blood count and LDH; imaging for symptoms.
- **Late effects:** anthracycline heart effects, neuropathy after BV or vincristine, radiation effects (nasal and sinus RT: dry mouth and eyes, hormone deficiencies), and transplant effects.
- **Infections:** vaccinate with non-live vaccines; HBV prophylaxis as planned; prompt treatment of fever.

When relapses happen: early and often in most PTCLs

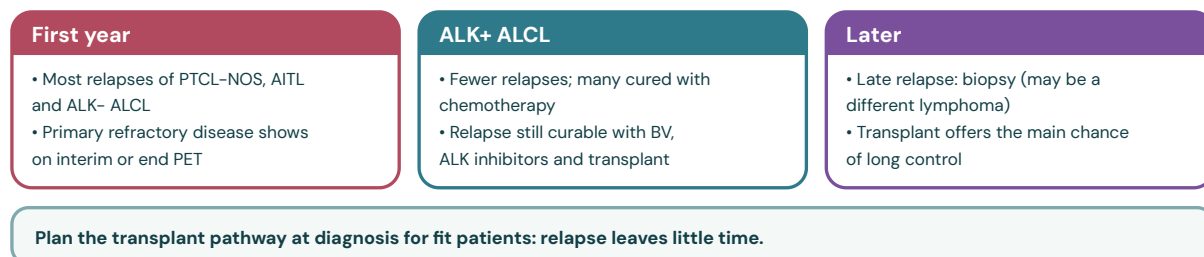


Figure 11. When relapses happen (UpToDate PTCL-NOS, initial and relapsed PTCL and sALCL reviews).

SHARE WITH PATIENTS: LIFE AFTER TREATMENT

Plain-language myth pages written for patients and families. Send them from the clinic to reinforce what you discussed.

[Lymphoma survivors can't return to normal life?](#)

[If the PET scan becomes negative, treatment can always be stopped immediately?](#)

09 Landmark trials

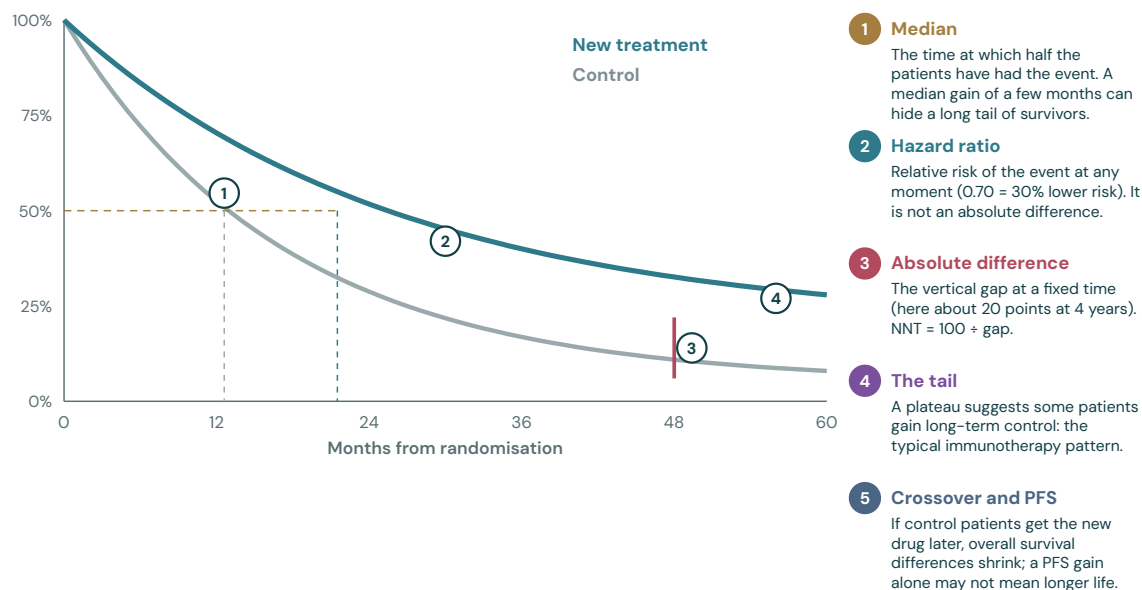
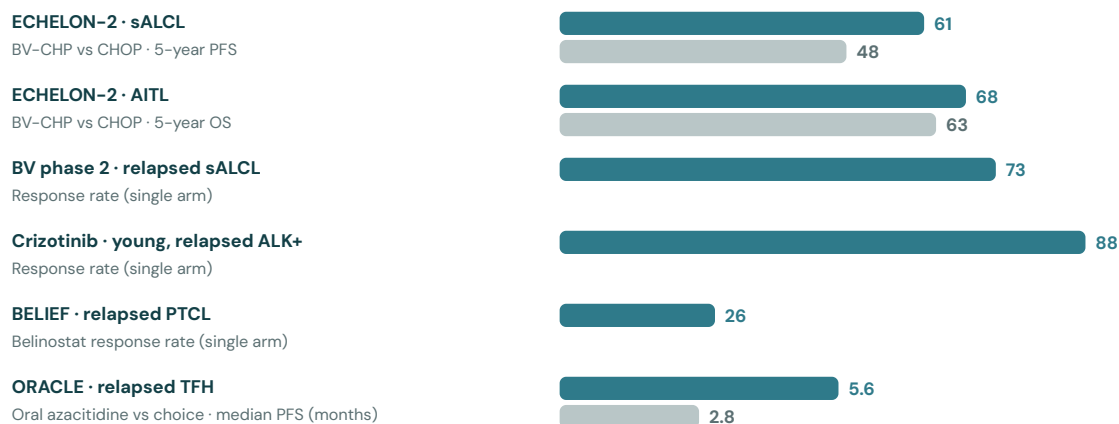


Figure 12. How to read a survival curve (illustrative). Use the absolute gap at a fixed time and overall survival, not the median or PFS alone, when weighing benefit against toxicity and cost.

The trials behind today's choices



Grey = comparator. Single-arm studies have no comparator bar. Endpoints differ: compare within a trial, not across trials.

Figure 13. Landmark results as reported in the UpToDate PTCL and sALCL reviews.

- **ECHELON-2**^[1]: BV-CHP vs CHOP in CD30-positive PTCL → better PFS and OS overall, driven by ALCL → BV-CHP standard for CD30-positive disease.
- **Swedish registry**^[9] and **DSHNHL**^[10]: etoposide helps younger patients.
- **NLG-T-01**^[11]: CHOEP then autologous transplant in first remission → the basis for consolidation in fit patients (no randomised proof).
- **BV phase 2**^[12]: high response rates in relapsed ALCL, some long remissions.
- **BELIEF**^[15] and **PROPEL**^[16]: modest single-agent activity in relapsed PTCL.
- **ORACLE**^[7]: oral azacitidine in relapsed TFH lymphoma → longer median PFS, endpoint not met.

10 Practice pearls & pitfalls

- A febrile patient with nodes, rash and high globulins may have AITL: biopsy a node rather than adding antibiotics or steroids.
- Ask for CD30 on every PTCL: at 1% or more, BV-CHP is the first-line choice.
- ALK-positive ALCL in a young patient is usually curable with chemotherapy alone; do not transplant in first remission.
- A necrotic nasal mass: deep biopsies and EBER; superficial necrotic tissue misses NK/T lymphoma.
- Hypercalcaemia with flower cells: test HTLV-1 (ATLL).
- Very high lymphocyte count with skin infiltration and effusions in an older adult: think T-PLL; ask for TCL1.
- Interim PET after three cycles: refractory disease needs a change of plan early.
- Plan transplant work-up during induction for fit patients with PTCL-NOS, AITL or ALK-negative ALCL.
- At relapse, biopsy: the histology may have changed, and EBV-driven B-cell lymphoma can arise in AITL.
- Screen for HLH (ferritin, triglycerides, fibrinogen) in a febrile, cytopenic patient with T-cell lymphoma.

11 Case vignettes: lines of management

Six cases from the clinic.

CASE 1 AITL · CD30 negative · fit

Fever, rash and nodes treated as typhoid

58 years, man

Stage IV

PIT 2

A 58-year-old shopkeeper has 6 weeks of fever, an itchy rash, generalised nodes and a palpable spleen. He received two courses of antibiotics for presumed typhoid. Haemoglobin 9.8 g/dL with a positive DAT; globulins high; LDH raised.

Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?

1

DIAGNOSE

Excision of an axillary node: T follicular helper lymphoma, angioimmunoblastic type (PD-1, ICOS, CXCL13 positive; CD21 meshworks; EBV-positive B blasts); CD30 under 1%.

2

STAGE

PET-CT: nodes above and below the diaphragm, spleen and marrow involved: stage IV; PIT 2 (raised LDH, marrow involvement).

3

TREAT

Fit and under 65: CHOEP × 6 with steroids for the haemolysis; PET after 3 cycles: partial response → complete six; end PET: complete response.

4

CONSOLIDATE

Fit, advanced, in CR: autologous transplant discussed (observation is an option; no randomised proof); he chooses transplant.

TEACHING POINTS

- AITL mimics infection and autoimmune disease: biopsy early.
- CD30 under 1%: CHOEP if fit; BV-CHP if CD30 positive.
- Decide consolidation at end-of-treatment PET.

See: Sections 02, 03 and 06, Algorithm 2.

CASE 2 ALK-positive ALCL · young

A 22-year-old with fast-growing neck nodes and bone pain

22 years, woman

Stage IV

ALK+

A 22-year-old student has neck nodes that doubled over 3 weeks, fever and pain in the right hip. LDH is raised.

Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?

1

DIAGNOSE

Excision: sheets of large hallmark cells, CD30 strong (membrane and Golgi), ALK positive (nuclear and cytoplasmic: NPM::ALK). Systemic ALK-positive ALCL.

2

STAGE

PET-CT: cervical and mediastinal nodes, right iliac bone: stage IV.

3

TREAT

BV-CHP × 6 (ECHELON-2); fertility counselling before treatment; interim PET complete response.

4

AFTER

End PET complete response: observation, no transplant. If she relapses: BV or crizotinib, then autologous transplant.

TEACHING POINTS

- ALK-positive ALCL: young, curable; BV-CHP first line.
- No transplant in first CR.
- Relapse is still treatable: BV, crizotinib, transplant.

See: Sections O4 and O6, Algorithm 3.

CASE 3 ALK-negative ALCL · relapse

Relapse four months after BV-CHP

61 years, man

Early relapse

BV-refractory

A 61-year-old with stage III ALK-negative ALCL achieved a partial response to BV-CHP. Four months later new nodes and fever appear.

Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?

1

BIOPSY

Node excision: ALK-negative ALCL again, CD30 strong.

2

CLASSIFY

Early (under 6 months) and BV-refractory relapse.

3

TREAT

Single-agent belinostat, pralatrexate or romidepsin rather than combination chemotherapy (UpToDate); PET after 3 cycles.

4

PLAN

If CR: autologous or allogeneic transplant in a fit patient; otherwise continue while benefiting; clinical trial preferred.

TEACHING POINTS

- Early or BV-refractory ALK-negative ALCL: newer single agents.
- Transplant is the only route to long-term control.
- Trials whenever possible.

See: Section O6 (relapse), Algorithm 4.

CASE 4 NK/T-cell lymphoma · nasal

A nasal ulcer called fungal sinusitis

44 years, man

Localised

EBER+

A 44-year-old farmer has 3 months of nasal blockage, foul discharge and a palatal ulcer. Two superficial biopsies showed "necrosis and acute inflammation"; antifungals did not help.

Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?

1 DIAGNOSE

Deep biopsy under anaesthesia: angiocentric infiltrate with necrosis; CD56, cytoplasmic CD3, granzyme positive; EBER positive. Extranodal NK/T-cell lymphoma, nasal type.

2 STAGE

MRI of nose and sinuses; PET-CT: disease confined to the nasal cavity and palate.

3 TREAT

Localised and fit: combined-modality treatment (asparaginase- or platinum-based chemotherapy with RT, concurrent or sandwich) at an experienced centre; plasma EBV DNA, endoscopy and PET to assess response; CR → observe.

4 TEACH

Superficial necrotic biopsies miss lymphoma; EBER is required for the diagnosis.

TEACHING POINTS

- Deep biopsy and EBER for every destructive midline lesion.
- No anthracycline regimens: asparaginase- or platinum-based chemotherapy with RT.
- Nasal NK/T lymphoma is seen in India more than in the West.

See: Sections 02, 03 and 06 (NK/T).

CASE 5 ATLL · hypercalcaemia**Confusion, high calcium and abnormal lymphocytes**

52 years, woman

Born in the Caribbean

HTLV-1+

A 52-year-old woman visiting family in Nellore is admitted with confusion, calcium 15 mg/dL, a white count of 38,000/μL with lobulated "flower" cells and a papular rash.

Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?

1 STABILISE

IV fluids and treatment of hypercalcaemia; check for Strongyloides before steroids.

2 DIAGNOSE

Flow cytometry: CD4+, CD25+, CD7-; HTLV-1 antibodies positive and confirmed: adult T-cell leukaemia-lymphoma, acute variant.

3 TREAT

Acute ATLL: VCAP-AMP-VECP (± mogamulizumab, avoided if allogeneic transplant is planned) or CHOP-like if unfit, with intrathecal prophylaxis; donor search for allogeneic transplant.

4 FAMILY

HTLV-1 counselling for partner and children (breastfeeding and sexual transmission).

TEACHING POINTS

- Hypercalcaemia with flower cells: ATLL until proven otherwise.
- HTLV-1 testing separates ATLL from PTCL and Sézary syndrome.
- Screen for Strongyloides.

See: Sections 02, 03 and 06 (ATLL).

CASE 6 PTCL-NOS · older

A 74-year-old with nodes and weight loss

74 years, man

Stage III

CD30 negative

A 74-year-old with diabetes and ischaemic heart disease (LVEF 50%) has 3 months of weight loss and generalised nodes.

Before you open the answer: what is the likely diagnosis, what must be done before treatment, and what is the plan?

1 **DIAGNOSE**

Excision: PTCL-NOS (CD3+, CD5 lost, CD30 negative; no TFH profile; EBER negative).

2 **TREAT**

Older, less fit, CD30-negative: CHOP × 6 with growth factor support; PET after 3 cycles.

3 **AFTER**

Complete response at the end: observation (not a transplant candidate); limited residual site: RT.

4 **AT RELAPSE**

Sequential single agents and palliative care; median survival after relapse without transplant is short.

TEACHING POINTS

- CHOP for older or less fit CD30-negative PTCL.
- RT for limited residual disease when transplant is not possible.
- Honest goals-of-care discussions at relapse.

See: Section 06, Algorithms 2 and 4.

12 References & guidelines

- NCCN Guidelines: T-Cell Lymphomas (current version); ESMO lymphoma guideline (2025); WHO Classification, 5th edition, and the International Consensus Classification (2022); Lugano classification (2014).
- Cross-checked in October 2026 against UpToDate topic reviews: PTCL–NOS diagnosis; T follicular helper cell lymphoma, angioimmunoblastic type; systemic ALCL diagnosis; extranodal NK/T-cell lymphoma diagnosis; ATLL diagnosis; T–PLL diagnosis; initial treatment of PTCL; initial treatment of systemic ALCL; relapsed or refractory PTCL; relapsed or refractory systemic ALCL; treatment of NK/T-cell lymphoma; treatment of ATLL; treatment of T–PLL. Citations from their reference lists. Indian data web–verified on 17 October 2026. Reviewed and approved by Dr. Haritha Chiramana, 17 October 2026.

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Educational summary for health professionals, written in our own words from current guidelines and primary trials. No drug doses are given. Hodgkin, aggressive B-cell and indolent B-cell lymphomas are parts 1–3 and cutaneous lymphomas part 5; leukaemias will follow as a separate module. Verify current guidelines, Indian regulatory approvals and individual patient factors before treatment decisions.