A PATIENT-PHYSICIAN COLLABORATIVE EFFORT

FOR A PATIENT FRIENDLY VERSION

TGCT Support, a program of the Life Raft Group (LRG), aims to improve the resources and outcomes for patients with tenosynovial giant cell tumor (TGCT), previously known as pigmented villonodular synovitis (PVNS). As patients carry the major burden of the disease, it is of the utmost importance that they have the ability to be active members of their healthcare team. This requires a shared language to overcome the medical jargon that often acts as a gatekeeper to patient involvement in their care. Therefore, a patient-physician collaborative effort led by TGCT Support, LRG, and SPAGN came together to create a patient-friendly version of the "Best Clinical Management Tenosynovial Giant Cell Tumour (TGCT): A consensus paper from the community of experts", enabling patients to participate in their care and inform their decision-making using the same evidence presented to clinicians, researchers, and other healthcare providers.

TGCT is comprised of two distinct subgroups, localized/nodular (N-TGCT) and diffuse TGCT (D-TGCT).

Nodular TGCT (N-TGCT) is well-defined in a single area of the joint and often rounded. N-TGCT is often cured by surgery alone and recurrence rates are low.

Diffuse TGCT (D-TGCT) is often extensive and involves the entirety of the joint and/or tendon sheath and may extend outside of the joint. D-TGCT has high recurrence and may be difficult to treat.

RECOMMENDATIONS*

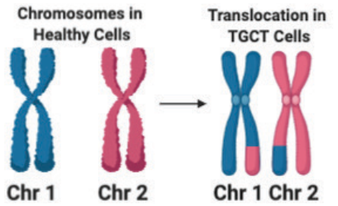
Diagnosis

MRI is the imaging technique of choice to detect recurrence.

Contrast dye, such as Gadolinium contrast, is recommended to allow with and without contrast dye subtraction.

Pathology

Under a microscope, N-TGCT and D-TGCT appear nearly identical. Both N-TGCT and D-TGCT are known for the over-production of the CSF1 protein caused by a gene abnormality, known as a chromosome translocation, only seen in 2-16% of tumour tissue.



Pathology review by an expert specialist is recommended, particularly in the case of discrepancies between imaging and symptoms.

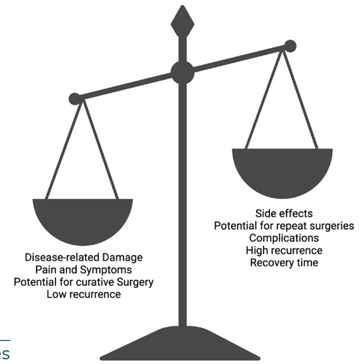
Treatment

Patients affected with TGCT should be treated within expert centers or reference networks, by a dedicated, experienced sarcoma multidisciplinary treatment team, including a pathologist, radiologist, orthopedic surgeon, pain specialist, surgical, radiation, and medical oncologists.

For suspected D-TGCT, a biopsy is recommended either by image-guided biopsy or during arthroscopic synovectomy (keyhole surgery).

Biopsy may be avoided if imaging is performed in an expert center, if the diagnosis is most obviously TGCT, and/or surgery is planned.

Active Monitoring



Active surveillance should be considered as the first option for asymptomatic patients. For symptomatic patients, active surveillance should also be considered if there is risk of major complications or suffering from surgery or medical treatment (e.g., history of severe toxicity from previous drug treatments, repeat operations and joint damage).

The decision to use active treatments such as surgery or medicine should balance the possibility of a cure, the impact the disease has on the patient, the risk of recurrence after surgery, and the potential for surgery-related complications and suffering.

When active surveillance is selected, the frequency of follow-up should be individualized based on tumour growth determined by MRI assessment, location of disease, and symptoms experienced by the patient.

Surgery

Surgical removal should be used when the disease is entirely capable of being removed and it can be accomplished without significant complications or long-term suffering, without recurrence, and if it improves quality of life.

*Recommendations are denoted in blue boxes

Radiation

The available literature provides insufficient evidence to propose a recommendation, thus we do not recommend the use of radiotherapy, either external beam and locally injected into the joint, as a standard treatment for TGCT.

Medications

Medicines that target the abnormal production of CSF1, a major driver of the growth of TGCT, are considered standard and a detailed list of available tumor response per medicine option is presented in the table.

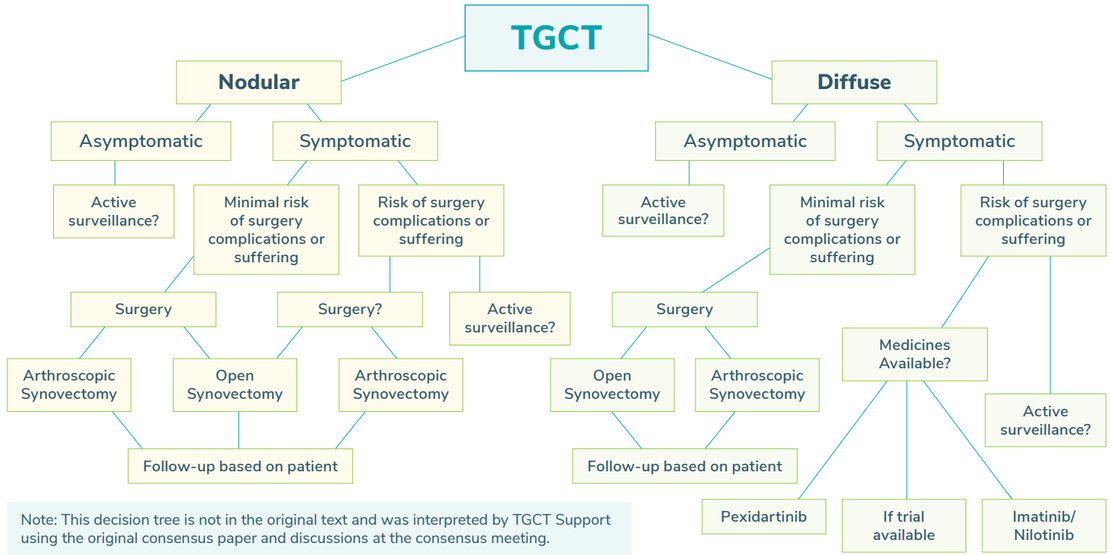
In countries without approved medicines, the preferred option should be clinical trial participation or off-label drugs that target CSF1 (e.g., imatinib, nilotinib) when available.

CURRENT MEDICINES AVAILABLE IN VARIOUS SETTINGS

MEDICINES	PERCENT OF PATIENTS THAT HAVE SIGNIFICANT SHRINKAGE (%)
Pexidartinib (Turalio)	39-62
Imatinib (Gleevec)	31
Nilotinib (Tasigna)	6
CLINICAL TRIAL MEDICINES	
Vimseltinib (DCC-3014)	49
Emactuzumab	71-85
Cabiralizumab	45

***AMB-05X has not published their trial results at this time and thus could not be included**

DECISION TREE ON POTENTIAL TREATMENTS



Note: This decision tree is not in the original text and was interpreted by TGCT Support using the original consensus paper and discussions at the consensus meeting.

TGCTSupport

www.tgctsupport.org

The mission of TGCT Support is to enhance treatment options and quality of life for TGCT patients through patient-powered research, education, empowerment, and global advocacy efforts.

READ MORE RECOMMENDATIONS HERE

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