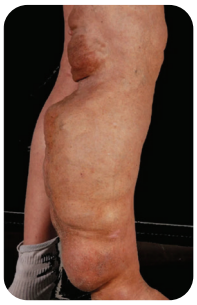
Leveraging ICD-10 Codes to Help Improve Care Quality and Outcomes in NF1-PN

Challenges With NF1-PN

- NF1 is an **autosomal dominant genetic disease** caused by variants in the NF1 gene, which encodes neurofibromin, a tumor suppressor protein[1]
- **Plexiform neurofibromas** are invasive nerve sheath tumors found in up to 50% of patients with NF1[2-4]
- NF1-PN impacts both adults and children, and is characterized by **debilitating morbidity, pain, disfigurement, and compression of internal organs**[1-5]

ICD-10 Codes for NF1-PN of the leg[6]:

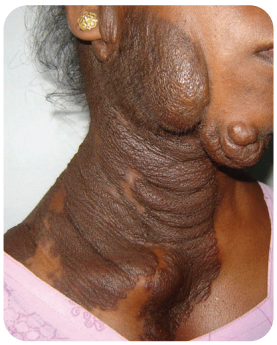


Q85.01:
Neurofibromatosis, type 1

Associated Plexiform Neurofibroma Code: D36.13

- Benign neoplasm of peripheral nerves and autonomic nervous system of lower limb, including hip

Capturing site-specific ICD-10-CM diagnosis codes for NF1-PN is critical for accurate diagnosis and to indicate patient- and disease-specific morbidities



ICD-10-CM for NF1-PN of the head/neck region[6]:

Q85.01:
Neurofibromatosis, type 1

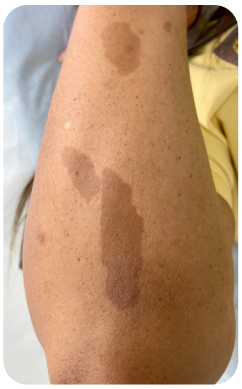
Associated Plexiform Neurofibroma Code: D36.11

- Benign neoplasm of peripheral nerves and autonomic nervous system of face, head, and neck

- Many patients with NF1-PN are diagnosed during childhood, but **are often lost to follow-up during adolescence or adulthood** due to limited treatment options and access to multidisciplinary care teams[7-10]
  - Insurance changes between adolescence and adulthood may disrupt access to NF1-PN treatment
  - **GOMEKLI® (mirdametinib) is the first FDA-approved systemic therapy for adults** with NF1-PN and is available in a formulation for patients with swallowing difficulties[11]

NF1-PN Is Characterized by Diagnostic Complexities

- Although clinical practice standards recommends that all patients diagnosed with NF1 (ICD-10-CM code Q85.01) be **assessed for the presence of plexiform neurofibromas**,[12] plexiform neurofibromas may be difficult to identify
  - **Misdiagnosis may lead to** inappropriate treatment decisions, delayed care, and adverse patient outcomes



Café-au-lait macules, a key diagnostic criterion for NF1[2,6]:

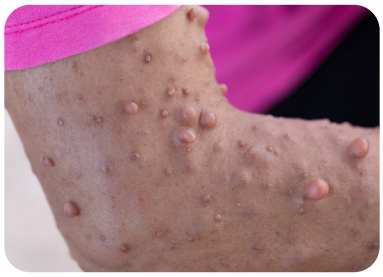
Associated Code for Café-au-lait macules: L81.3

- Café-au-lait spots

Café-au-lait macules are pigmentary skin features that often present prior to infancy in patients with NF1[1]

Screening for key signals of NF1 including café-au-lait macules or cutaneous neurofibromas[2,13] is critical to support accurate diagnosis

Cutaneous neurofibromas are found in nearly all patients with NF1 during their lifetime[6,13]:



Associated Code for Cutaneous Neurofibromas: D23.9

- Other benign neoplasm of skin, unspecified

Cutaneous neurofibromas are complications of the skin, often present during adolescence, and are associated with itching, disfigurement, and pain[1]

INDICATION

GOMEKLI (mirdametinib) is indicated for the treatment of adult and pediatric patients 2 years of age and older with neurofibromatosis type 1 (NF1) who have symptomatic plexiform neurofibromas (PN) not amenable to complete resection.

IMPORTANT SAFETY INFORMATION

GOMEKLI Warnings and Precautions are Ocular Toxicity, Left Ventricular Dysfunction, Dermatologic Adverse Reactions, and Embryo-Fetal Toxicity.

ICD-10-CM=*International Classification of Diseases, Tenth Revision, Clinical Modification*; NF1-PN=neurofibromatosis type 1-associated plexiform neurofibromas.

Please see additional Important Safety Information on page 4 and click here for full Prescribing Information.

SlicerDicer: A Critical NF1-PN Patient Identification Tool

- Ensure your EHR is prepared for re-engaging patients. SlicerDicer is **a reporting tool that enables healthcare teams** to visualize patient population data to assess specific groups based on diagnosis, lab results, demographics, and other clinical criteria
  - Those data can be modified or filtered to include/remove criteria, and can be displayed as a summary, detailed patient list, or chart
- The clinical criteria used to generate the report must be searchable within the electronic health record
- **SlicerDicer has the ability to:**
  - Simplify access to complex clinical and operational data by transforming it into digestible and actionable information to support better care
  - Support the identification of NF1-PN patient population lost to follow up
  - Identify patients who may have been misdiagnosed or underdiagnosed
  - Examine trends in outcomes and access to inform decision making and efficiency

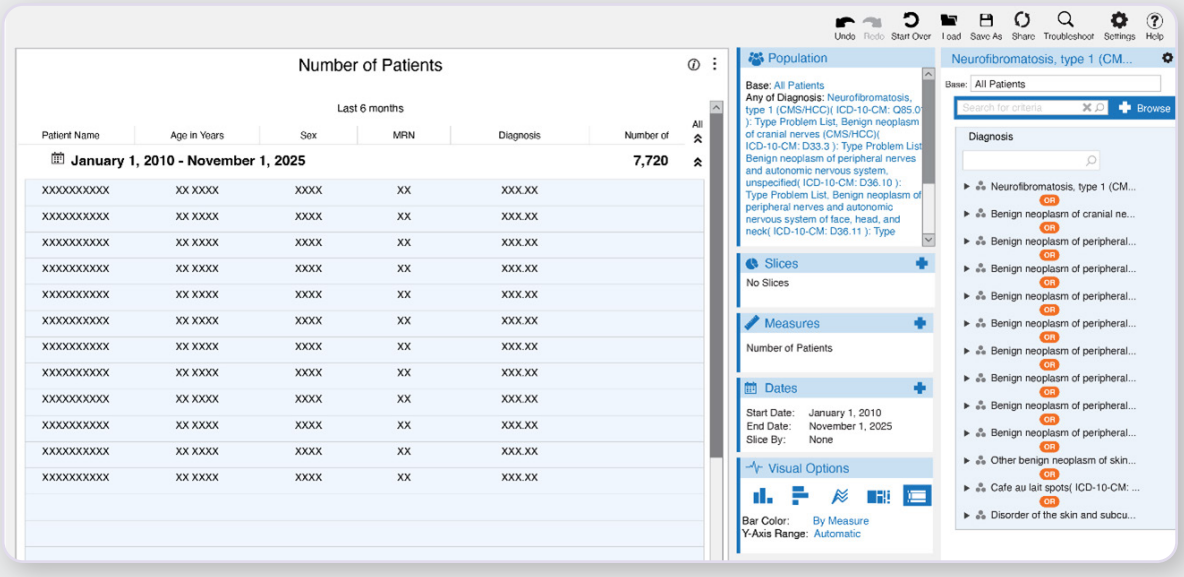
- The instructions for SlicerDicer are not intended to provide guidance or recommendations concerning treatment. The user and their health system are solely responsible for implementation, testing, and monitoring of the instructions to the right. While EHRs may assist providers in identifying appropriate patients, prescribing decisions are the sole responsibility of the provider

Using SlicerDicer in Epic to Identify Patients With NF1-PN or Patients Who May Need Further Evaluation

1. Access **SlicerDicer** by clicking the **Epic logo > Reports > SlicerDicer**
2. Select **Patients** Data Model
3. Set appropriate **Base**, for example, **All Patients**
4. Type **Diagnosis** in the **+ Browse** search bar and:
   - **a.** If an NF1-PN Diagnosis Grouper is available, select the **Diagnosis by grouper** criteria, and select the **NF1-PN** Diagnosis Grouper
   - **b.** If an NF1-PN Diagnosis Grouper is not available, select the **Diagnosis** criteria, enter **ICD-10-CM codes from page 4 in the search field**
5. Select appropriate **Measures**, for example, **Number of Patients *(default)***
6. Select appropriate **Dates**, for example, **Last 12 months**
7. Select appropriate **Visual Options**, for example, **Detail Table and Cross Tab**
8. Click **Save As**
9. In the Create New Session window:
   - **a.** **Name** as appropriate, for example, **Patients with NF1-PN**
   - **b.** Add an appropriate explanation within the **Description** section to define what the report depicts
   - **c.** Set appropriate lookback range using **Start Date** and **End Date**, and assign a **Time Period**, for example, **12 months**
   - **d.** Based on users' permissions, this report can be saved as a **Public** or **Private** session
10. Click **Create Session**

Re-engaging previously disconnected patients is essential to providing the care they need

SlicerDicer in Action



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Improving NF1-PN Diagnostics and Outcomes With ICD-10 Codes

Using the ICD-10 codes listed below and the steps presented for the SlicerDicer reporting tool may help:

- Ensure underserved patients receive care according to clinical guidelines and care team expertise
- Prepare you to invite patients back for reassessment to optimize their care by reducing delays and closing potential gaps in care
- Coordinate the multidisciplinary care team, which may include geneticists, oncologists, neurologists, and other specialists

ICD-10-CM Code[6]	Description
NF1	
Q85.01	Neurofibromatosis, type 1
NF1-associated PNs	
D33.3	Benign neoplasm of cranial nerves
D36.10	Benign neoplasm of peripheral nerves and autonomic nervous system, unspecified
D36.11	Benign neoplasm of peripheral nerves and autonomic nervous system of face, head, and neck
D36.12	Benign neoplasm of peripheral nerves and autonomic nervous system, upper limb, including shoulder
D36.13	Benign neoplasm of peripheral nerves and autonomic nervous system of lower limb, including hip
D36.14	Benign neoplasm of peripheral nerves and autonomic nervous system of thorax
D36.15	Benign neoplasm of peripheral nerves and autonomic nervous system of abdomen
D36.16	Benign neoplasm of peripheral nerves and autonomic nervous system of pelvis
D36.17	Benign neoplasm of peripheral nerves and autonomic nervous system of trunk, unspecified

ICD-10 Codes Consistent With NF1-PN Findings

The following ICD-10-CM codes describe clinical manifestations that may be present in patients with NF1. Therefore, these codes may be indicative of misdiagnosis or underdiagnoses of NF1-PN

ICD-10-CM Code[6]	Description
D23.9	Other benign neoplasm of skin, unspecified (typically used to code for cutaneous neurofibromas)
L81.3	Café-au-lait spots
L98.9	Disorder of the skin and subcutaneous tissue (typically used to code for axillary or inguinal skinfold freckling)
H21.89	Other specified disorders of iris and ciliary body (typically used to code for lich nodule on the eye)
D17.9	Benign lipomatous neoplasm, unspecified (typically used to code for lipomas)
L91.8	Other hypertrophic disorders of the skin (typically used to code for skin tags)

Accurate documentation of diagnoses allows for broader recognition of NF1-PN, helps to raise disease awareness, and enables identification of appropriate patients to support patient care

CLICK HERE to discover platform-specific EHR guides with proposed steps to create an order set and help streamline other processes for the NF1-PN care team

Please see Important Safety Information on page 4 and click here for full Prescribing Information.

INDICATION AND IMPORTANT SAFETY INFORMATION

INDICATION

GOMEKLI (mirdametinib) is indicated for the treatment of adult and pediatric patients 2 years of age and older with neurofibromatosis type 1 (NF1) who have symptomatic plexiform neurofibromas (PN) not amenable to complete resection.

WARNINGS AND PRECAUTIONS

Ocular Toxicity: GOMEKLI can cause ocular toxicity including retinal vein occlusion (RVO), retinal pigment epithelium detachment (RPED), and blurred vision. In the adult pooled safety population, ocular toxicity occurred in 28% of patients treated with GOMEKLI: 21% were Grade 1, 5% were Grade 2 and 1.3% were Grade 3. RVO occurred in 2.7%, RPED occurred in 1.3%, and blurred vision occurred in 9% of adult patients. In the pediatric pooled safety population, ocular toxicity occurred in 19% of patients: 17% were Grade 1 and 1.7% were Grade 2. Conduct comprehensive ophthalmic assessments prior to initiating GOMEKLI, at regular intervals during treatment, and to evaluate any new or worsening visual changes such as blurred vision. Continue, withhold, reduce the dose, or permanently discontinue GOMEKLI as clinically indicated.

Left Ventricular Dysfunction: GOMEKLI can cause left ventricular dysfunction. GOMEKLI has not been studied in patients with a history of clinically significant cardiac disease or LVEF <55% prior to initiation of treatment. In the ReNeu study, decreased LVEF of 10 to <20% occurred in 16% of adult patients treated with GOMEKLI. Five patients (9%) required dose interruption, one patient (1.7%) required a dose reduction, and one patient required permanent discontinuation of GOMEKLI. The median time to first onset of decreased LVEF in adult patients was 70 days. Decreased LVEF of 10 to <20% occurred in 25%, and decreased LVEF of ≥20% occurred in 1.8% of pediatric patients treated with GOMEKLI. One patient (1.8%) required dose interruption of GOMEKLI. The median time to first onset of decreased LVEF in pediatric patients was 132 days. All patients with decreased LVEF were identified during routine echocardiography, and decreased LVEF resolved in 75% of patients. Before initiating GOMEKLI, assess ejection fraction (EF) by echocardiogram. Monitor EF every 3 months during the first year and then as clinically indicated. Withhold, reduce the dose, or permanently discontinue GOMEKLI based on severity of adverse reaction.

Dermatologic Adverse Reactions: GOMEKLI can cause dermatologic adverse reactions including rash. The most frequent rashes included dermatitis acneiform, rash, eczema, maculo-papular rash and pustular rash. In the pooled adult safety population, rash occurred in 92% of patients treated with GOMEKLI (37% were Grade 2 and 8% were Grade 3) and resulted in permanent discontinuation in 11% of patients. In the pooled pediatric safety population, rash occurred in 72% of patients treated with GOMEKLI (22% were Grade 2 and 3.4% were Grade 3) and resulted in permanent discontinuation in 3.4% of patients. Initiate supportive care at first signs of dermatologic adverse reactions. Withhold, reduce the dose, or permanently discontinue GOMEKLI based on severity of adverse reaction.

Embryo-Fetal Toxicity: GOMEKLI can cause fetal harm when administered to a pregnant woman. Verify the pregnancy status of females of reproductive potential prior to the initiation of GOMEKLI. Advise pregnant women and females of reproductive potential of the potential risk to a fetus. Also advise patients to use effective contraception during treatment with GOMEKLI and for 6 weeks after the last dose (females) or 3 months after the last dose (males).

ADVERSE REACTIONS

The most common adverse reactions (>25%) in adult patients were rash (90%), diarrhea (59%), nausea (52%), musculoskeletal pain (41%), vomiting (38%), and fatigue (29%). Serious adverse reactions occurred in 17% of adult patients who received GOMEKLI. The most common Grade 3 or 4 laboratory abnormality (>2%) was increased creatine phosphokinase.

The most common adverse reactions (>25%) in pediatric patients were rash (73%), diarrhea (55%), musculoskeletal pain (41%), abdominal pain (39%), vomiting (39%), headache (34%), paronychia (32%), left ventricular dysfunction (27%), and nausea (27%). Serious adverse reactions occurred in 14% of pediatric patients who received GOMEKLI. The most common Grade 3 or 4 laboratory abnormalities (>2%) were decreased neutrophil count and increased creatine phosphokinase.

USE IN SPECIFIC POPULATIONS

Verify the pregnancy status of patients of reproductive potential prior to initiating GOMEKLI. Due to the potential for adverse reactions in a breastfed child, advise patients not to breastfeed during treatment with GOMEKLI and for 1 week after the last dose.

To report SUSPECTED ADVERSE REACTIONS, contact SpringWorks Therapeutics Inc. at 1-888-400-7989 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

Please click here for full Prescribing Information including Patient Information and Instructions for Use.

References: 1. Gutmann DH et al. *Nat Rev Dis Primers*. 2017;3:17004. **2.** Miller DT et al. *Pediatrics*. 2019;143(5):e20190660. **3.** Prada CE et al. *J Pediatr*. 2012;160(3):461-467. **4.** Ejerskov C et al. *Oncol Ther*. 2023;11(1):97-110. **5.** Darrigo LG Jr et al. *Brain Behav*. 2022;12(6):e2599. **6.** ICD-10-CM files. Centers for Disease Control and Prevention. Accessed September 10, 2025. https://www.cdc.gov/nchs/icd/icd-10-cm/files.html **7.** Radtke HB et al. *Pediatric Health Med Ther*. 2023;14:19-32. **8.** Rietman AB et al. *Am J Med Genet A*. 2018;176(5):1150-1160. **9.** Foji S et al. *Health Expect*. 2022;25(2):659-666. **10.** Data on file. Trinity market research. **11.** GOMEKLI. Prescribing Information. SpringWorks Therapeutics, Inc. **12.** Fisher MJ et al. *Neuro Oncol*. 2022;24(11):1827-1844. **13.** Pulh P et al. *Acta Neuropathol Commun*. 2025;13:158.

CLICK HERE to learn more about GOMEKLI.

SpringWorks THERAPEUTICS

GOMEKLI® (mirdametinib)
1 mg tablets for oral suspension
1 mg and 2 mg capsules