Nervous System Tumor Susceptibility Syndromes: From Pathophysiology to New Treatments

Jaishri Blakeley, MD
The Marjorie Bloomberg Tiven Professor of Neurofibromatosis
Professor of Neurology, Neurosurgery and Oncology
Johns Hopkins University
Baltimore, Maryland, USA

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- **Advisory Boards:**
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  - Springworks
  - Medical Advisory Board: Children's Tumor Foundation

- **Off label drugs**:
  - Bevacizumab
  - Brigatinib
  - Neratinib
  - Binimetinib

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Learning Objectives

- Awareness of the genetic causes and clinical presentation of the most common tumor susceptibility syndromes associated with neoplasms of the nervous system:
  - Lynch Syndrome
  - The Neurofibromatoses:
    - Neurofibromatosis Type 1
    - Neurofibromatosis Type 2-Schwannomatosis
    - Other Schwannomatosis
  - Li-Fraumeni Syndrome
  - Tuberous Sclerosis

- Awareness of major therapeutic developments for NF1, NF2-SWN and Tuberous Sclerosis.

Key Message

- Tumor susceptibility syndromes have well defined genetic causes and molecular targets that have led to some therapeutic breakthroughs.

- Although rare overall, these syndromes are associated with many common nervous system tumor and non-tumor manifestations (including neurovascular, neurocognitive, epilepsy, and neuropathy manifestations). Improved pathophysiologic understanding of these conditions (and the associated therapeutic advances) has the potential to benefit both the people living with these genetic conditions as well as the broader population living with neurologic morbidities.