Why discuss mesenchymal tumour syndromes?

- Young patients
- Sarcoma survivors at increased risk of second cancers
- Sarcomas overrepresented among survivors of melanoma, breast cancer, thyroid cancer, Hodgkin lymphoma and leukemia
- Rare genetic syndromes often associated with mesenchymal tumors:
  - Li Fraumeni, Retinoblastoma, Rothmund Thompson syndrome
  - Neurofibromatosis type 1
  - Familial adenomatous polyposis
  - Carney-Stratakis
  - Hereditary leiomyomatosis and renal cell cancer

Disclosure of Relevant Financial Relationships

USCAP requires that all planners (Education Committee) in a position to influence or control the content of CME disclose any relevant financial relationship WITH COMMERCIAL INTERESTS which they or their spouse/partner have, or have had, within the past 12 months, which relates to the content of this educational activity and creates a conflict of interest.

Dr. Bovée declares she has no conflict(s) of interest to disclose.

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Dr. Bovée declares he/she has no conflict(s) of interest to disclose.
Are sarcomas hereditary?

Thus, when patients ask if their sarcomas are hereditary, the answer is now: “Yes, at least partly, in most cases”.

Benjamin and Futreal, Lancet Oncol 2016

Non-hereditary tumour syndromes

- Somatic mosaic conditions
- Rather common for mesenchymal tumors:
  - McCune Albright / polyostotic fibrous dysplasia GNAS
  - Ollier disease / Maffucci syndrome IDH

Role of the pathologist

- Identify these patients
  - Based on combination of different tumours
  - Based on specific morphology
- Recommend genetic counseling
- Include this information at multidisciplinary tumour boards:
  - Surveillance and prevention
  - Therapy

THANK YOU