

INTRODUCTION

- **BAP1-TPDS** is a genetic autosomal dominant inheritance condition characterized by mutations in the BAP1 gene
- located on chromosome 3p21.1, encodes a deubiquitinating enzyme involved in DNA repair, cell cycle regulation, and chromatin remodeling.
- Loss of function leads to genomic instability and increased cancer susceptibility across multiple tissue types.
- Associated malignancies include malignant mesothelioma, uveal melanoma, cutaneous melanoma, renal cell carcinoma, basal cell carcinoma
- Early detection of mesothelioma is critical due to poor prognosis and late-stage diagnoses in most patients.
- The NIH Clinical Center has implemented a comprehensive surveillance protocol for early mesothelioma detection in BAP1-TPDS patients that integrates a multidisciplinary perioperative approach.

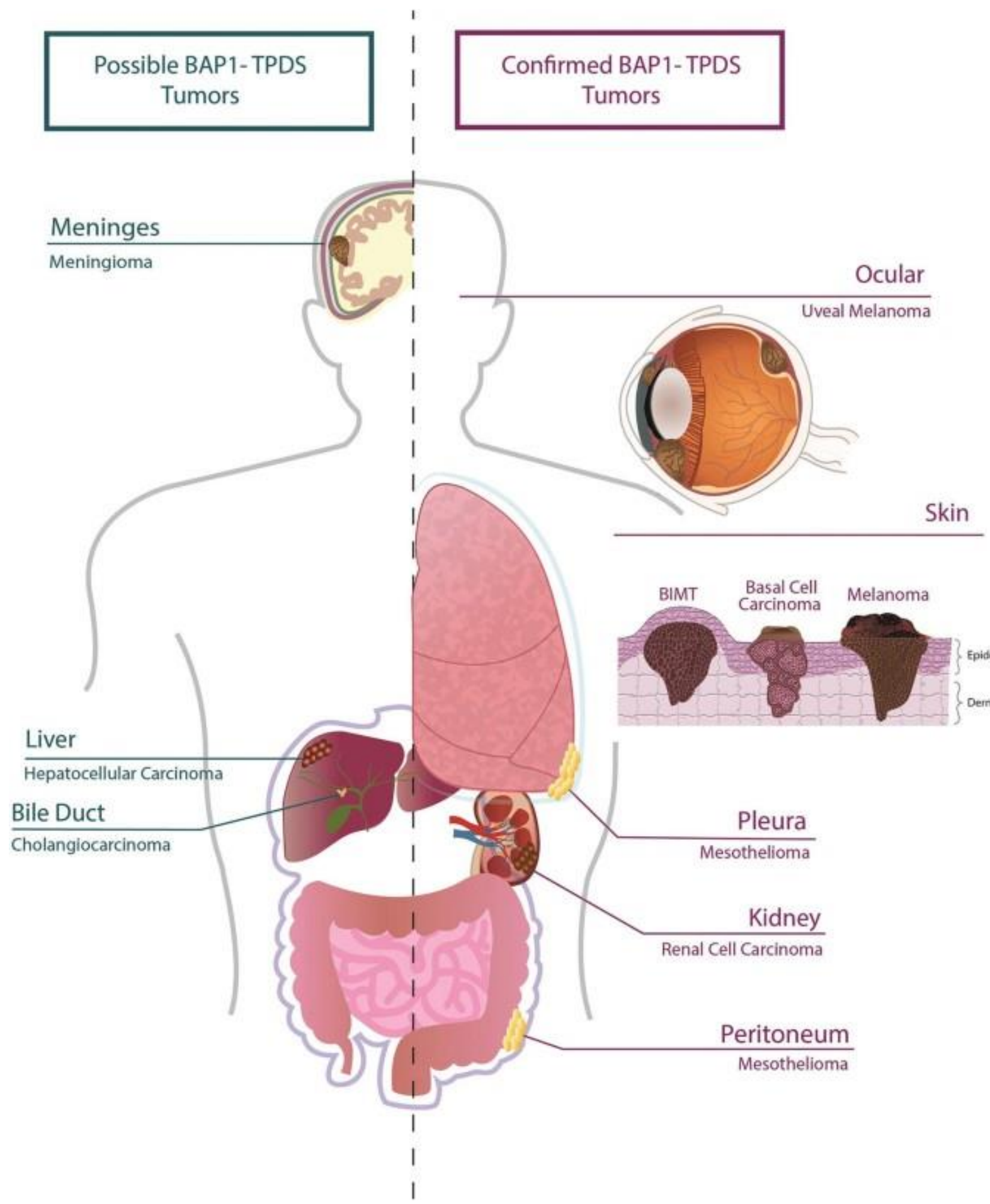


Image 1: BAP1-TPDS impacted organs.

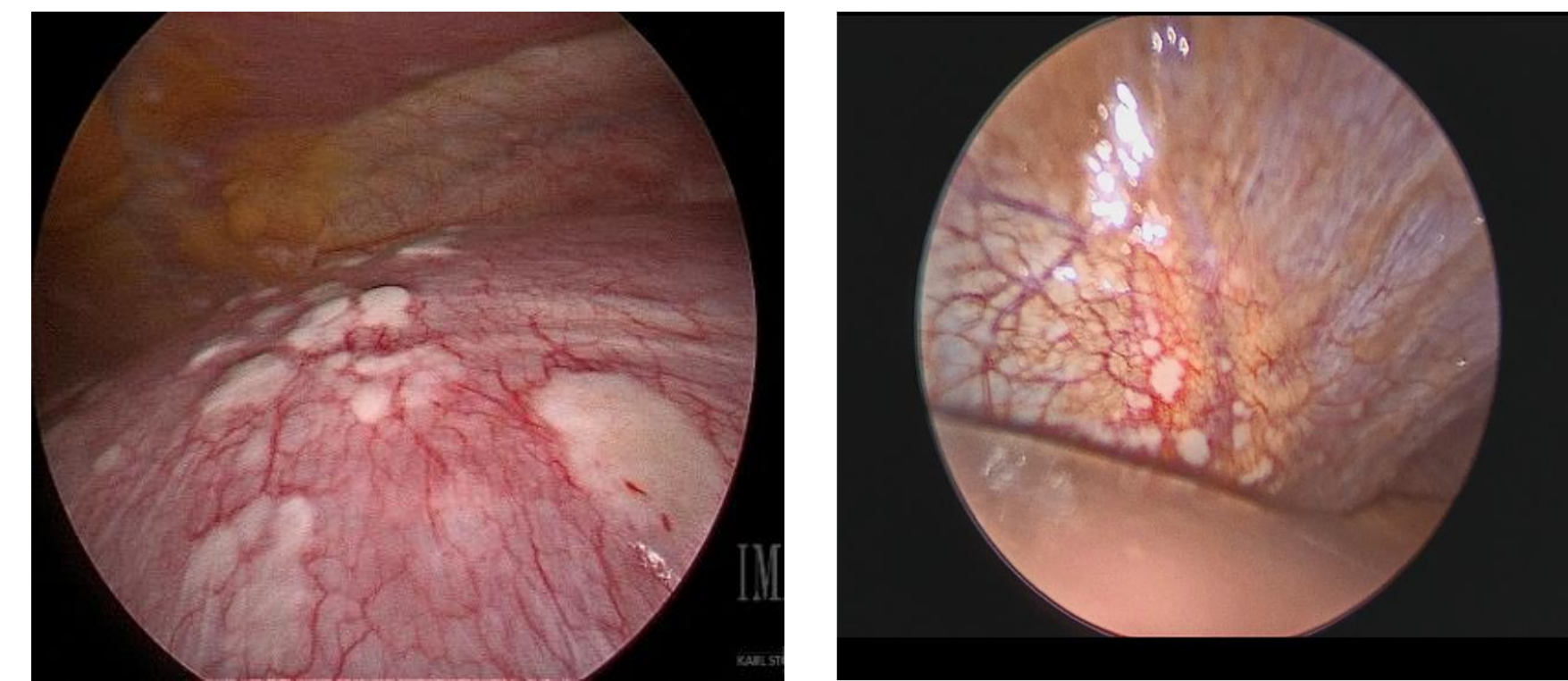


Image 2: left, early MPM in thoracic cavity; right, early MPM in peritoneal cavity

DIAGNOSIS, SURVEILLANCE, AND TREATMENT

- **Dual-energy CT (DECT)** imaging is used to detect early pleural and peritoneal mesothelioma in high-risk individuals with germline BAP1 mutations.
- **Liquid biopsy (cfDNA)** to monitor for circulating tumor DNA and identify mesothelioma-related genetic markers even before clinical symptoms appear.
- Minimally invasive procedures performed in a single operative session
 - **Bronchoscopy:** for evaluating potential lung involvement, particularly for early pleural mesothelioma detection
 - **Bilateral Video Assisted Thoracic Surgery (VATS):** allows for thorough assessment of thoracic involvement, including lung biopsies
 - **Diagnostic Laparoscopy:** evaluate the abdominal cavity and assess peritoneal mesothelioma
 - **Cystoscopy:** evaluation of urothelial involvement in cases of renal cell carcinoma or other urological malignancies

SURGICAL SET-UP



Images A-D Surgical Set-up
A: Bronchoscopy
B: Diagnostic Laparoscopy
C: Cystoscopy
D: VATs

INTRAOPERATIVE CONSIDERATIONS

Positioning



Image 3: lateral position for VATS



Image 4: supine position for diagnostic laparoscopy & cystoscopy (male patients)



Image 5: frog-leg position for cystoscopy (female patients)

Maintaining Sterility

- Aseptic technique maintained throughout the multi-procedure case.
- Each procedure treated as a separate sterile event.
- Instruments and supplies opened only as needed.
- Surgical field re-prepped and re-draped between procedures/anatomical sites.
- Contaminated equipment removed promptly from the sterile field.
- Gloves and gowns changed as indicated.
- OR traffic minimized and team communication maintained to preserve sterility

INTRAOPERATIVE CONSIDERATIONS CONT.

Multidisciplinary Perioperative Coordination

- Preoperative planning conducted with surgery, anesthesia, nursing, and ancillary services to coordinate multiple procedures.
- Procedure sequence confirmed to optimize efficiency and safety.
- Equipment, instrumentation, and room setup coordinated in advance for each surgical specialty.
- Anesthesia management adjusted to support procedure transitions and patient positioning.
- Clear communication maintained during handoffs and between procedural phases.
- Roles and responsibilities defined for each team to ensure smooth workflow.
- Sustained collaboration facilitated efficient transitions while preserving standards of patient safety and sterility.

SPECIMEN MANAGEMENT

- Specimens collected, labeled, and handled according to policy.
- Specimen type, source, and laterality verified with the surgical team.
- Separate specimens maintained and clearly identified for each procedure.
- Proper containment used to prevent contamination or loss.
- Specimens promptly transferred to pathology with required documentation.
- Research specimen procurement practices maintained to preserve research data

For Right /Left VATs

- Right/Left Skin Biopsy - Research
- Right/Left Diaphragm and Chest Wall – Frozen/Path
- Right/Left Normal Pleura – Research

For Diagnostic Laparoscopy

- Right/Left Peritoneal Surface Biopsy - Path

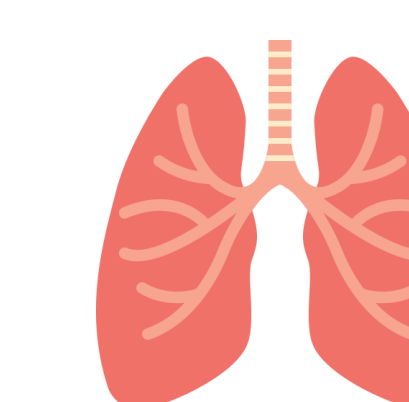
For Cystoscopy

- Urine specimen for research
- Additional biopsies if needed

REFERENCES



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