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**Introduction & Objectives:** Primary urethral cancer (PUC) is a rare cancer entity. Due to the low incidence of this malignancy, the main body of literature consists mainly of case reports, making evidence-based management recommendations difficult. We aim to review reported disease management strategies of PUC and their impact on oncologic outcomes.

**Materials & Methods:** A systematic research was performed using Medline, Scopus and Web of Science, according to the PRISMA statement, to find studies of the last ten years with more than 20 patients investigating treatment strategies and their differential impact on outcomes.

**Results:** In localized PUC, penis-sparing surgery can be performed in male patients, while in females, complete urethrectomy with surrounding tissue is advised to minimize recurrence due to positive margins. Radiotherapy (RT) has worse survival and recurrence rates than surgery, as well as adverse effects, limiting its current use in genital preserving therapy.

Locally advanced PUC should be treated with multimodal therapy (MMT), as monotherapies result in inferior recurrence and survival rates. Extent of surgery is still undecided, favoring radical cysto (prostate) urethrectomy (RCU). Lymph node involvement is a predictor for survival, highlighting the role of lymph node dissection for disease control and staging. Radiotherapy (RT) can improve survival in combination with surgery and/or chemotherapy (CHT). Neoadjuvant platin-based CHT can improve overall survival and recurrence-free survival.

At recurrence, salvage therapy with surgery and/or CHT can improve survival.

Superficial urothelial carcinoma of the prostatic urethra can be treated with transurethral resection, while stromal invasion often features concomitant bladder cancer with poor prognosis require RCU with or without systemic preoperative chemotherapy. Metastatic PUC is treated in analogy to metastatic urothelial carcinoma.

**Conclusions:** PUC is a rare malignancy with often a poor natural history, requiring a stage- and gender-specific risk-based treatment strategy. The role of systematic perioperative chemotherapy and extent of surgery is becoming important.