

ICUD guidelines for upper tract urothelial carcinoma: a state-of-the-art evidence-based guidance for clinical decisions regarding diagnosis, management and treatment

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Upper tract urothelial carcinoma (UTUC) accounts for 5% of all urothelial carcinomas with an estimated annual incidence of 1–2 cases per 100,000 inhabitants. Despite a common histologic appearance, epidemiologic, molecular, embryologic/anatomic (urogenital sinus versus ureteric bud), practical/therapeutic, clinico-pathologic and prognostic differences suggest that UTUC and bladder urothelial carcinoma (UC) represent two disparate disease entities [1]. UTUCs, for example, are more often invasive than bladder UC at diagnosis suggesting a differentially more aggressive phenotype [2]. We, indeed, now know that we cannot extrapolate what we know from bladder UC unequivocally to UTUC.

This realization has led to an acceleration in the research which collided with a significant progress in technological innovations necessary for the diagnosis and management of UTUC. Indeed, we have seen an insurgence of interest in UTUC by scientists, clinicians and the industry alike. This, in turn, has led to a better understanding of the biology underlying the natural history of UTUC thereby ushering the age of personalized medicine in this remote disease.

To help make sense of the explosion of data in UTUC, the ICUD and SIU have partnered up to create modern clinical guidelines. The main aim was to identify, summarize

and evaluate the highest quality evidence and most current data about prevention, diagnosis, risk stratification, prognosis, and therapy. To this end, we brought the most knowledgeable experts in this field worldwide together, defined the most important questions related to clinical practice in UTUC and identified all possible decision options and their outcomes. This guideline integrates the identified decision points and respective courses of action with the clinical judgment and experience of practitioners.

The mission of this guideline is to improve our understanding and eventually care of a disease that is greatly understudied and underappreciated. Special focus of this effort was to integrate the newest data regarding susceptibility and causative factors, contemporary concepts and controversies in the diagnosis and staging, prediction tools and their value in decision-making within each disease stage as well as patient selection and treatment options.

In addition to the well-established association with smoking exposure, aristolochic acid and defects in mismatch repair genes associated with Lynch syndrome have been identified as UTUC-specific risk factors. A growing understanding of other biological pathways involved in UTUC has unveiled prognostic/predictive biomarkers that promise to improve our risk stratification to increase accuracy thereby facilitating individualized treatment strategies.

We further cover hot topics such as the selection and approach to kidney-sparing management to avoid overtreatment, the different approaches to radical nephroureterectomy, distal ureterectomy, and lymphadenectomy with their differential impact on outcomes. Further covered areas by this comprehensive body of work are the role of post-surgery intravesical instillation and, not to be underestimated, systemic chemotherapy.

While these guidelines present a reliable, up-to-date integrative help for clinicians, the low incidence of the

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disease still limits the quality and, therefore, the degree of the recommendations. Indeed, despite growing interest and understanding of UTUC, its management remains challenging, requiring further high-quality multicenter collaborations/clinical trials with a prospective design.

Foremost international experts in UTUC have come together from different countries and specialties to inform us on the newest evidence that will shape our practice tomorrow. I am deeply grateful to them and to my co-chairs for relentlessly pushing towards excellence and nothing less. We hope that these reports of the ICUD/SIU guidelines will be of great value to you.

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