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NEPHRON-SPARRING SURGERY FOR UNILATERAL SYNDROMIC WILM'S TUMOR: OUR EXPERIENCE.

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Aim: Wilms tumor (WT) is one of the most common pediatric renal malignancies. Currently, radical nephroureterectomy is the procedure of choice for non-syndromic unilateral WT. However, bilateral tumors and unilateral WT with genetic predisposition and certain risk factors, are strong indications for nephron-sparing surgery (NSS). In recent years, NSS has also gained interest in anatomically favorable unilateral tumors that meet certain criteria.

Case report: We present two cases of unilateral syndromic WT. Both patients were diagnosed with WT1-related Wilms Tumor Syndrome and PAX8 gene mutation. They both received neoadjuvant chemotherapy. Upon therapy completion, both patients showed favorable overall clinical and paraclinical status hence, NSS with enucleation and marginal resection was performed. Postoperative histopathology results revealed complete tumor resection in the first patient and small missed lesions on the surgical margins of the second which pointed out the need for possible repeat nephron-sparing surgery. Intraoperatively, we decided to perform a complete kidney resection, as further NSS would leave insufficient renal parenchyma for proper renal function.

Conclusion: NSS is the preferred procedure for unilateral WT with genetic predisposition, due to high risk of asynchronous contralateral tumor development. There is ongoing debate regarding the advancement of the method in patients with no genetic predisposition but anatomically favorable tumors after neoadjuvant chemo. Repeat NSS could be considered when remnant small lesions, local relapse or metachronous tumor development

occur with satisfactory results on the renal function, depending on the location and size of the affected parenchyma. Therefore, detailed, and individualized evaluation and planning of each case is essential.