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Uretero-Inguinal Hernia

Uretero-inguinal hernia (UIH) is an exceedingly rare condition, characterized by the displacement of the ureter into the inguinal canal. This phenomenon can be congenital or acquired and is often associated with complex anatomical anomalies or predisposing factors. It poses diagnostic and therapeutic challenges due to its unusual presentation and the potential for severe complications if not identified and managed appropriately.

UIH has two primary classifications: paraperitoneal and extraperitoneal. Paraperitoneal UIH, which constitutes approximately 80% of cases, involves the ureter adhering to a hernial sac and being pulled into the inguinal canal. This type is often linked to sliding hernias and may involve other abdominal viscera. In contrast, extraperitoneal UIH, accounting for the remaining 20%, occurs without a hernial sac and is typically associated with congenital anomalies of the ureteral and renal systems. This variety is believed to result from abnormal embryological development, such as late separation of the Wolffian duct or adherence of the ureter to genitoinguinal structures.

UIH predominantly affects males, possibly due to the developmental descent of the Wolffian duct structures into the scrotum, creating a pathway for ureteral involvement. In adults, risk factors include advanced age, obesity, renal transplantation, and collagen disorders. In children, the condition is exceptionally rare, with only a limited number of documented cases.

Clinical manifestations of UIH are varied and depend on the extent of ureteral involvement and the presence of secondary complications. Patients may present with symptoms ranging from an asymptomatic inguinal mass to signs of obstructive uropathy, such as flank pain, hematuria, or hydronephrosis. In many cases, UIH is discovered incidentally during surgical exploration for inguinal hernia repair. Imaging modalities like ultrasound, computed tomography (CT), and voiding cystourethrography (VCUG) play critical roles in preoperative diagnosis, helping identify ureteral involvement and associated urinary tract anomalies.

Management of UIH requires careful surgical intervention to prevent iatrogenic injuries. The approach varies depending on the type and severity of the hernia, as well as the patient's overall condition. For paraperitoneal UIH, high ligation of the hernial sac and repositioning of the ureter are common strategies. For extraperitoneal cases, interventions may include ureteral reimplantation or ureteroneocystostomy, especially in the presence of significant obstruction or stricture.

In pediatric cases, the rarity of UIH necessitates heightened clinical awareness, particularly in the presence of congenital urological anomalies. Early recognition and intervention are essential to avoid complications like ureteral injury or progressive renal impairment. Long-

term follow-up with renal function tests and imaging is crucial to monitor outcomes and prevent recurrence.

The literature highlights the importance of individualized care and the role of multidisciplinary teams, including pediatric surgeons, urologists, and radiologists, in managing this complex condition. Advances in laparoscopic techniques have improved visualization and allowed for more precise interventions, reducing morbidity, and enhancing recovery.

UIH represents a fascinating interplay between congenital and acquired factors, with implications for both surgical practice and urological management. Continued documentation of cases and research into the underlying mechanisms will be essential to refine diagnostic and therapeutic strategies for this rare entity.

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Antithrombotic Therapy

Antithrombotic therapy for children is a rapidly evolving area of medical research and practice due to the increasing recognition and diagnosis of thromboembolic events (TEs) in pediatric populations. Unlike adults, children experience TEs primarily as a consequence of severe illness or medical interventions, such as central venous catheterization. This distinct etiology necessitates tailored approaches to diagnosis, treatment, and prevention.

Pediatric TEs differ significantly from adult cases in terms of epidemiology, pathophysiology, and therapeutic implications. While the incidence of VTE in the general pediatric population remains low (0.07 to 0.14 per 10,000 children), hospitalized children face a much higher risk—up to 1000-fold greater—due to the widespread use of central venous access devices (CVADs) and other invasive procedures. Neonates and adolescents constitute the most vulnerable groups, reflecting distinct physiological and pathophysiological factors such as immature coagulation systems and pubertal hormonal changes.

The pediatric coagulation system undergoes significant maturation during the first year of life, which alters the pharmacodynamics and pharmacokinetics of anticoagulant

medications. For instance, younger children often require higher weight-based doses of anticoagulants, despite having lower levels of coagulation proteins. These differences pose unique challenges in drug selection, dosing, and monitoring.

Historically, antithrombotic therapy in children has relied on unfractionated heparin (UFH), low-molecular-weight heparin (LMWH), and vitamin K antagonists (VKAs). However, recent advances have introduced direct oral anticoagulants (DOACs) as a promising alternative due to their consistent pharmacokinetics, ease of administration, and reduced monitoring requirements. Clinical trials have demonstrated that DOACs, such as rivaroxaban and dabigatran, are as effective as standard anticoagulants while offering improved safety profiles.

The American Society of Hematology (ASH) 2018 guidelines emphasize the use of anticoagulation in symptomatic VTE and stress the importance of individualized therapy based on the patient's clinical status and risk factors. For asymptomatic cases, the decision to treat remains contentious, reflecting the low certainty of evidence regarding the balance between risks and benefits. The recommendations also underline the need for multidisciplinary care involving pediatric hematologists to optimize treatment outcomes.

Updated guidance from the International Society on Thrombosis and Haemostasis (ISTH) has refined outcome definitions for pediatric VTE clinical trials, introducing parameters like patient-important bleeding to standardize safety assessments. These developments aim to enhance the comparability and applicability of trial results.

Despite significant progress, challenges remain. Many recommendations for pediatric antithrombotic therapy are extrapolated from adult studies due to the limited number of pediatric-specific trials. This reliance underscores the need for robust, age-appropriate research to address gaps in knowledge, particularly regarding long-term outcomes and the management of chronic conditions such as post-thrombotic syndrome.

Meta-analyses and network comparisons have further clarified the efficacy and safety of various anticoagulants. For example, DOACs have shown non-inferiority to traditional agents in preventing recurrent TEs, with lower risks of major bleeding. However, concerns persist about their use in specific pediatric subgroups, such as neonates and critically ill children, highlighting the importance of cautious implementation based on individual risk profiles.

Prophylactic anticoagulation remains a debated topic in pediatric care. Although standard in adult practice, its routine use in children is not widely endorsed due to the scarcity of high-quality evidence supporting its benefits. Studies investigating the role of prophylaxis in high-risk settings, such as CVAD-related thrombosis, have yielded mixed results, further complicating clinical decision-making.

Emerging research continues to expand the therapeutic arsenal for pediatric TEs. The advent of age-specific formulations of DOACs, coupled with advances in imaging and biomarker technologies, holds promise for improving diagnostic precision and treatment

efficacy. Moreover, ongoing trials are expected to address critical questions about optimal dosing, duration of therapy, and long-term safety.

In conclusion, antithrombotic therapy in children has evolved significantly, driven by a growing understanding of pediatric hemostasis and advances in pharmacology. While traditional anticoagulants remain the cornerstone of treatment, DOACs represent a paradigm shift in managing pediatric TEs. Nevertheless, the field requires continued investment in research and collaboration to refine therapeutic strategies and ensure the best outcomes for young patients.

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Adamkiewicz Artery in Pediatric Posterior Thoracic Tumors

Posterior thoracic tumors in pediatric patients, particularly those of neurogenic origin such as neuroblastomas and ganglioneuromas, pose unique surgical challenges due to their proximity to the Artery of Adamkiewicz (AKA). This artery, the primary blood supply to the anterior spinal cord, is often located between the T8 and L1 vertebral levels and exhibits significant anatomical variability. Inadvertent injury to the AKA during tumor resection can lead to catastrophic outcomes, including anterior spinal cord ischemia, paraparesis, or paraplegia. Consequently, accurate preoperative identification of the AKA is critical to surgical planning and patient safety.

Recent advancements in imaging techniques, particularly spinal angiography (SA) and Magnetic Resonance Angiography (MRA), have revolutionized preoperative evaluation for pediatric patients with posterior thoracic tumors. These modalities allow detailed mapping of the spinal vasculature, enabling surgeons to plan resections that minimize the risk of vascular injury. Studies have demonstrated that incorporating these imaging techniques into preoperative protocols not only improves surgical outcomes but also reduces the incidence of neurologic complications.

One study involving 36 pediatric patients evaluated the utility of preoperative spinal angiography. Among these patients, SA identified the AKA in all cases, demonstrating its

reliability in mapping vascular anatomy. In four cases where the AKA was in close proximity to the tumor, surgical plans were modified, with three patients undergoing non-surgical management, such as radiation therapy, to mitigate risk. Importantly, no complications arose from the SA procedure itself, highlighting its safety and efficacy as a preoperative tool.

Another key finding from recent research is the significant reduction in neurologic complications when SA is employed. A retrospective analysis comparing outcomes before and after the routine use of SA in pediatric posterior thoracic tumor resections revealed a marked decrease in the incidence of postoperative spinal ischemia. Prior to implementing routine SA, one patient in the study cohort developed paraplegia following resection. Post-SA implementation, no such complications were observed, underscoring the role of detailed preoperative vascular mapping in enhancing patient safety.

In addition to spinal angiography, Magnetic Resonance Angiography (MRA) has shown promise as a non-invasive alternative for visualizing the AKA. A case report highlighted the successful use of MRA in a 14-month-old child with a thoracic neuroblastoma. The imaging identified the precise location of the AKA, allowing the surgical team to avoid critical vascular structures during resection. MRA's non-invasive nature and ability to provide high-resolution images make it particularly suitable for pediatric patients, where minimizing procedural risks is paramount.

Despite these advancements, challenges remain in achieving consistent and accurate preoperative identification of the AKA. The artery's variability in origin, pathway, and laterality necessitates a tailored approach for each patient. Moreover, the choice of imaging modality—whether SA, MRA, or a combination—often depends on institutional resources and expertise. While spinal angiography remains the gold standard for AKA visualization, its invasive nature, and associated risks, though minimal, must be carefully weighed against the benefits in each case.

The integration of imaging findings into surgical decision-making has profound implications for treatment strategies. In cases where the AKA is identified in close proximity to the tumor, surgeons may opt for partial resections, alternative surgical approaches, or adjunctive therapies such as radiation. This tailored approach not only preserves spinal cord function but also improves overall outcomes by reducing the likelihood of tumor recurrence or residual disease.

Studies also emphasize the importance of interdisciplinary collaboration in managing these complex cases. The involvement of pediatric surgeons, interventional radiologists, and neuro-oncologists ensures a comprehensive evaluation of risks and benefits, facilitating informed decision-making. Multidisciplinary tumor boards play a pivotal role in this process, integrating imaging findings with clinical and pathological data to devise individualized treatment plans.

Further research is needed to refine imaging techniques and establish standardized protocols for preoperative evaluation of the AKA. Emerging technologies, such as advanced MRI sequences and 3D vascular mapping, hold promise for enhancing the accuracy and accessibility of preoperative imaging. Additionally, longitudinal studies assessing long-term outcomes in patients undergoing surgery with preoperative AKA identification will provide valuable insights into the efficacy of these strategies.

In conclusion, the preoperative identification of the Adamkiewicz Artery is a critical component of surgical planning for pediatric posterior thoracic tumors. Techniques such as spinal angiography and Magnetic Resonance Angiography enable precise vascular mapping, significantly reducing the risk of spinal ischemia and associated neurologic complications. By incorporating these modalities into preoperative protocols and fostering interdisciplinary collaboration, healthcare teams can optimize surgical outcomes and improve the quality of life for pediatric patients with these challenging tumors. Continued advancements in imaging technology and research will further enhance the safety and efficacy of these interventions, paving the way for improved standards of care in pediatric oncology.

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