PEDIATRIC SURGERY *Update**
Volume 63 No. 04 OCTOBER 2024

Laparoscopic Retroperitoneal Lymph Node Dissection

Laparoscopic Retroperitoneal Lymph Node Dissection (RPLND) is a crucial surgical technique primarily used in the management of testicular cancer and certain pediatric malignancies such as paratesticular rhabdomyosarcoma. Over the years, the technique has evolved significantly with advancements in minimally invasive surgery and the advent of robotic assistance, offering improved outcomes and reduced morbidity for patients. This review synthesizes information from multiple studies to provide a comprehensive understanding of the efficacy, safety, and advancements in laparoscopic and robot-assisted RPLND.

The earliest use of laparoscopic RPLND was reported in 1992, marking a significant shift from the traditional open surgical approach. The primary advantage of laparoscopic RPLND over open surgery lies in its minimally invasive nature, which translates to reduced postoperative pain, shorter hospital stays, and quicker recovery times. A study from 2010 highlighted the successful use of laparoscopic RPLND in high-risk pediatric patients with paratesticular rhabdomyosarcoma (PTRMS). The study reported that laparoscopic RPLND is a safe and effective diagnostic and therapeutic procedure for children over ten years of age with primary tumors larger than 5 cm. The average operative time was 382 minutes, with minimal blood loss and no significant postoperative complications, allowing for rapid commencement of adjuvant chemotherapy.

Robot-assisted RPLND (RA-RPLND) emerged as a significant advancement in the early 2000s, offering enhanced precision and visualization through robotic technology. The da Vinci Surgical System, for instance, has been instrumental in performing complex dissections with improved dexterity and control. A 2017 study documented the use of the da Vinci Xi system for RA-RPLND in adolescent patients. The study included two cases: a 17-year-old male with a mixed non-seminomatous germ cell tumor and a 15-year-old male with ectomesenchymoma. Both cases reported successful outcomes with no intraoperative complications and minimal blood loss. The robotic approach allowed for precise nerve-sparing dissections, preserving ejaculatory function in the patients.

Further supporting the efficacy of RA-RPLND, a 2012 study presented two adolescent cases involving paratesticular rhabdomyosarcoma (PT-RMS) and testicular germ cell tumor (T-GCT). The study emphasized that RA-RPLND is not only feasible but also provides excellent oncologic outcomes with low morbidity. The enhanced three-dimensional visualization and precise instrumentation afforded by robotic systems contribute to better

surgical outcomes, including reduced risk of complications such as vascular and bowel injuries.

Despite the advantages, the learning curve associated with RA-RPLND is steep, requiring significant expertise in both laparoscopic and robotic surgery. A comprehensive review of robotic-assisted surgeries highlighted that proficiency in RA-RPLND could take up to 200 cases. However, the benefits, including reduced operative times and enhanced safety through robotic training modules, justify the initial learning curve. Additionally, the use of proctors and robotic training modules can further mitigate the risks associated with the learning curve, ensuring safer and more efficient surgeries.

Long-term oncologic outcomes of RA-RPLND have been encouraging. Studies have shown that RA-RPLND offers comparable, if not superior, results to traditional open and laparoscopic approaches in terms of disease-free survival rates. For instance, patients undergoing RA-RPLND for clinical stage I non-seminomatous germ cell tumors have reported high safety and early oncologic effectiveness, with minimal long-term complications. Moreover, the minimally invasive nature of the procedure reduces the likelihood of postoperative complications such as chylous ascites, ileus, and small bowel obstruction, which are more common in open surgeries.

The role of RA-RPLND in pediatric populations, although limited, has shown promising results. Pediatric patients, due to their smaller anatomical structures, present unique challenges in surgical management. The precision and enhanced visualization provided by robotic systems are particularly beneficial in this demographic, allowing for meticulous dissection and preservation of vital structures. Studies have reported that RA-RPLND in pediatric patients results in shorter hospital stays, quicker recovery times, and lower incidence of complications compared to open surgery.

However, despite its advantages, RA-RPLND is not without its limitations. The high cost of robotic systems and the need for extensive training are significant barriers to widespread adoption. Additionally, the lack of long-term data in pediatric populations necessitates further research to fully understand the long-term outcomes and potential late effects of the procedure. Ongoing studies and patient registries are crucial in addressing these gaps and providing more comprehensive data on the efficacy and safety of RA-RPLND.

In conclusion, laparoscopic and robot-assisted RPLND represent significant advancements in the surgical management of retroperitoneal lymph node dissections. The transition from open surgery to minimally invasive and robotic-assisted techniques has resulted in improved patient outcomes, reduced morbidity, and quicker recovery times. While the learning curve and high costs associated with robotic systems remain challenges, the benefits offered by RA-RPLND, particularly in terms of precision and safety, make it a valuable option in the surgical armamentarium. Continued research and long-term studies will further elucidate the role of RA-RPLND in both adult and pediatric populations, ensuring that patients receive the most effective and least invasive treatment options available.

References:
1- Tomaszewski JJ, Sweeney DD, Kavoussi LR, Ost MC: Laparoscopic retroperitoneal lymph node dissection for high-risk pediatric patients with paratesticular rhabdomyosarcoma. J Endourol. 24(1):31-4, 2010
2- Cost NG, DaJusta DG, Granberg CF, Cooksey RM, Laborde CE, Wickiser JE, Gargollo PC: Robot-assisted laparoscopic retroperitoneal lymph node dissection in an adolescent population. J Endourol. 26(6):635-40, 2012
3- Glaser AP, Bowen DK, Lindgren BW, Meeks JJ: Robot-assisted retroperitoneal lymph node dissection (RA-RPLND) in the adolescent population. J Pediatr Urol. 13(2):223-224, 2017
4- Mansfield SA, Murphy AJ, Talbot L, Prajapati H, Maller V, Pappo A, Singhal S, Krasin MJ, Davidoff AM, Abdelhafeez A: Alternative approaches to retroperitoneal lymph node dissection for paratesticular rhabdomyosarcoma. J Pediatr Surg. 55(12):2677-2681, 2020
5- Brown CT, Sebasti?o YV, Zann A, McLeod DJ, DaJusta D: Utilization of robotics for retroperitoneal lymph-node dissection in pediatric and non-pediatric hospitals. J Robot Surg. 14(6):865-870, 2020
6- Li W, Xiong L, Zhu Q, Lu H, Zhong M, Liang M, Jiang W, Wang Y, Cheng W: Assessment of retroperitoneal lymph node status in locally advanced cervical cancer. BMC Cancer. 21(1):484, 2021

Anaplastic Thyroid Cancer

Anaplastic Thyroid Cancer (ATC) is an exceedingly rare and aggressive form of thyroid malignancy. It accounts for less than 2% of all thyroid cancers but is responsible for a disproportionate number of thyroid cancer deaths due to its rapid progression and poor prognosis.

Anaplastic Thyroid Cancer predominantly affects older adults, typically presenting in the sixth to seventh decade of life. However, there are rare instances of ATC in younger populations, including children. According to a 2018 case report, ATC in children is extremely uncommon, with one of the youngest reported cases being a five-year-old child. Most thyroid malignancies in younger patients are well-differentiated papillary and follicular variants, with ATC being a rare progression from these types.

The clinical presentation of ATC often includes a rapidly enlarging neck mass, pain, dysphagia, and symptoms related to airway obstruction. The aggressive nature of the tumor leads to early metastasis, primarily to the lungs, bones, and brain. Diagnostic imaging such as CT and MRI are essential in evaluating the extent of local invasion and distant metastasis. A case reported in 2018 described a young child with a rapidly progressing thyroid mass that necessitated an emergency tracheostomy.

Histologically, ATC is characterized by a high degree of cellular atypia and the absence of the typical nuclear features seen in differentiated thyroid cancers. DICER1 mutations are increasingly recognized in poorly differentiated thyroid carcinoma, a precursor to ATC, particularly in younger patients. Immunohistochemical staining plays a crucial role in the diagnosis, with markers such as EMA and cytokeratin being positive in ATC cells, while markers like TTF-1 and thyroglobulin are usually negative.

Molecular studies have identified several genetic mutations associated with ATC. The BRAFT1799A mutation is common, particularly in cases arising from previously differentiated thyroid cancers. Other mutations frequently seen include TP53 and TERT

promoter mutations. The identification of these mutations has implications for targeted therapy, although the aggressive nature of ATC often limits the effectiveness of such treatments.

The management of ATC is challenging due to its aggressive nature and poor response to conventional therapies. Surgery, when feasible, is the primary treatment modality. Total thyroidectomy is recommended to achieve local control, although the extensive local invasion often limits the completeness of surgical resection. Adjuvant therapies, including radiation and chemotherapy, have limited efficacy.

Recent advances in molecular biology have opened new avenues for targeted therapies. Inhibitors targeting specific genetic mutations, such as BRAF and MEK inhibitors, have shown promise in early studies. Immunotherapy, particularly checkpoint inhibitors, is also being investigated as a potential treatment option for ATC. However, the rarity of the disease poses significant challenges in conducting large-scale clinical trials to validate these approaches.

The prognosis for ATC remains poor, with a median survival of less than six months from diagnosis. Early diagnosis and aggressive treatment are critical to improving outcomes. Multimodal therapy, including surgery, radiation, and chemotherapy, may offer some benefit in locoregionally confined disease. Patients who received aggressive treatment had a median survival of 13 months, compared to less than six months for those who received less aggressive management.

In conclusion, Anaplastic Thyroid Cancer is a highly aggressive malignancy with a poor prognosis. The rarity of the disease, particularly in younger patients, underscores the need for increased awareness and early diagnosis. Advances in molecular genetics offer hope for targeted therapies, although their clinical utility remains to be fully established. Continued research and clinical trials are essential to develop more effective treatments and improve survival outcomes for patients with this devastating disease.

References:

1- Wu H, Sun Y, Ye H, Yang S, Lee SL, de las Morenas A: Anaplastic thyroid cancer: outcome and the mutation/expression profiles of potential targets. Pathol Oncol Res. 21(3):695-701, 2015 2- Sharma SC, Sakthivel P, Raveendran S, Singh CA, Nakra T, Agarwal S: Anaplastic Carcinoma Thyroid in a Young Child - an Extremely Rare Occurrence. Acta Medica (Hradec Kralove). 61(4):150-152, 2018

3- Caperton CO, Jolly LA, Massoll N, Bauer AJ, Franco AT: Development of Novel Follicular Thyroid Cancer Models Which Progress to Poorly Differentiated and Anaplastic Thyroid Cancer. Cancers (Basel). 13(5):1094, 2021

4- Xu B, David J, Dogan S, Landa I, Katabi N, Saliba M, Khimraj A, Sherman EJ, Tuttle RM, Tallini G, Ganly I, Fagin JA, Ghossein RA: Primary high-grade non-anaplastic thyroid carcinoma: a retrospective study of 364 cases. Histopathology. 80(2):322-337, 2022

5- Gunda V, Ghosh C, Hu J, Zhang L, Zhang YQ, Shen M, Kebebew E: Combination BRAFV600E Inhibition with the Multitargeting Tyrosine Kinase Inhibitor Axitinib Shows Additive Anticancer Activity in BRAFV600E-Mutant Anaplastic Thyroid Cancer. Thyroid. 33(10):1201-1214, 2023

6- Ver Berne J, Van den Bruel A, Vermeire S, De Paepe P: DICER1 Mutations Define the Landscape of Poorly Differentiated Thyroid Carcinoma in Children and Young Adults: Case Report and Literature Review.

Am J Surg Pathol. 2024 Jun 24. doi: 10.1097/PAS.0000000000002265. Epub ahead of print. PMID: 38912716.

US for Necrotizing Enterocolitis

Ultrasound has increasingly become a vital tool in diagnosing and managing necrotizing enterocolitis (NEC) in neonates, particularly premature infants. NEC is a significant cause of morbidity and mortality in this vulnerable population, and early, accurate diagnosis is crucial for effective treatment. Traditional abdominal radiographs (KUB) have long been the standard for diagnosing NEC, but their limitations have led to the exploration of ultrasound as a complementary or alternative imaging modality.

One of the primary advantages of ultrasound over radiography is its ability to provide real-time, dynamic imaging of the bowel and surrounding structures. This capability allows for the assessment of bowel wall thickness, perfusion, and the presence of free fluid or air in the abdomen, which are critical indicators of NEC progression. Moreover, ultrasound can detect pneumatosis intestinalis, portal venous gas, and pneumoperitoneum with higher sensitivity than radiographs, especially in the early stages of NEC.

Radiographs, while useful, have significant limitations in sensitivity, often failing to detect early signs of NEC. Studies have shown that radiographs may miss up to 50% of early NEC cases, as their sensitivity ranges from 13% to 25% for detecting key features such as free air, which is identified in only 44% of perforated NEC cases. This underlines the importance of more sensitive imaging techniques, such as ultrasound, which can identify subtle changes in bowel perfusion and wall thickness before they become apparent on radiographs.

The technical aspects of performing abdominal ultrasound for NEC diagnosis are well-documented. Techniques such as gray-scale and Doppler ultrasound are employed to evaluate bowel wall integrity and blood flow, respectively. For instance, intramural gas (pneumatosis) appears as echogenic dots within the bowel wall, while portal venous gas shows up as echogenic particles in the liver parenchyma. These features are often detectable earlier and more clearly with ultrasound than with radiographs.

Several studies have highlighted the diagnostic and prognostic value of ultrasound in NEC. In a prospective study by Dordelmann et al., routine ultrasound screening of premature infants identified portal venous gas as a specific indicator of NEC, absent in other clinical conditions. Similarly, a meta-analysis by Cuna et al. demonstrated that ultrasound features such as free air, absent peristalsis, and complex ascites were strongly associated with the need for surgical intervention or death.

Despite its advantages, the use of ultrasound for NEC diagnosis is not without challenges. Overlying bowel gas can obscure underlying structures, making it difficult to obtain clear images. Additionally, the skill and experience of the sonographer and radiologist play a crucial role in accurately interpreting ultrasound findings. There is also variability in the

availability of ultrasound in neonatal intensive care units (NICUs), particularly outside regular hours, which can limit its use in urgent situations.

The integration of ultrasound into NEC diagnosis protocols offers several benefits. For example, it can expedite the diagnosis, allowing for earlier intervention and potentially reducing the severity of the disease and its complications. Ultrasound also aids in stratifying patients who may fail medical management and require surgical intervention. This stratification is crucial as the outcomes for surgically treated NEC are generally poorer compared to medically managed cases.

Further research and quality improvement studies are needed to establish standardized protocols for the use of ultrasound in NEC. Advances in ultrasound technology, such as contrast-enhanced ultrasound, hold promise for even more accurate detection of bowel perfusion and other critical parameters. Such advancements could significantly improve the sensitivity and specificity of ultrasound, making it an even more valuable tool in the fight against NEC.

In conclusion, while abdominal radiographs remain the standard imaging modality for NEC diagnosis, the addition of ultrasound can enhance diagnostic accuracy and patient outcomes. The real-time, detailed imaging capabilities of ultrasound provide critical insights into bowel health and disease progression, offering a more comprehensive approach to managing NEC. Continued research, training, and technological advancements are essential to fully integrate ultrasound into standard NEC diagnostic protocols, ultimately improving care for neonates affected by this severe condition.

References:

1- Cuna AC, Reddy N, Robinson AL, Chan SS. Bowel ultrasound for predicting surgical management of necrotizing enterocolitis: a systematic review and meta-analysis. *Pediatr Radiol*. 48(5):658-666, 2018

2- Raghuveer TS, Lakhotia R, Bloom BT, Desilet-Dobbs DA, Zarchan AM. Abdominal Ultrasound and Abdominal Radiograph to Diagnose Necrotizing Enterocolitis in Extremely Preterm Infants. *Kans J Med*. 26;12(1):24-27, 2019

3- Chan B, Gordon S, Yang M, Weekes J, Dance L. Abdominal Ultrasound Assists the Diagnosis and Management of Necrotizing Enterocolitis. *Adv Neonatal Care*. 21(5):365-370, 2021

4- Hwang M, Tierradentro-García LO, Dennis RA, Anupindi SA. The role of ultrasound in necrotizing enterocolitis. *Pediatr Radiol*. 52(4):702-715, 2022

5- Cuna A, Chan S, Jones J, Sien M, Robinson A, Rao K, Opfer E. Feasibility and acceptability of a diagnostic randomized clinical trial of bowel ultrasound in infants with suspected necrotizing enterocolitis. *Eur J Pediatr*. 181(8):3211-3215, 2022

6- May LA, Epelman M, Daneman A. Ultrasound for necrotizing enterocolitis: how can we optimize imaging and what are the most critical findings? *Pediatr Radiol*. 53(7):1237-1247, 2023

7- Kallis MP, Roberts B, Aronowitz D, Shi Y, Lipskar AM, Amodio JB, Aggarwal A, Sathya C. Utilizing ultrasound in suspected necrotizing enterocolitis with equivocal radiographic findings. *BMC Pediatr*. 23(1):134, 2023

8- May LA, Costa J, Hossain J, Epelman M. The role of an abbreviated ultrasound in the evaluation of necrotizing enterocolitis. *Pediatr Radiol*. 54(6):944-953, 2024

***Edited by: Humberto Lugo-Vicente, MD, FACS, FAAP**

Professor of Pediatric Surgery, UPR - School of Medicine, UCC School of Medicine & Ponce School of Medicine.
Pediatric Surgery, San Jorge Children's & Woman Hospital.
Postal Address: P.O. Box 10426, San Juan, Puerto Rico USA 00922-0426.
Tel (787) 340-1868 E-mail: *pediatricsurgerypr@gmail.net*
Internet: pedsurgeryupdate.com

**** PSU 1993-2024***
ISSN 1089-7739