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Lymphatic Duct Embolization

Lymphatic duct embolization (LDE) has emerged as a significant intervention for pediatric patients suffering from complex lymphatic disorders. This procedure, which involves blocking abnormal lymphatic vessels, is particularly beneficial in treating conditions like chylothorax, Kaposi form lymphangiomatosis (KLA), and other central lymphatic flow disorders. The following discussion delves into the clinical applications, methodologies, outcomes, and advancements in LDE for children, drawing from ten detailed studies.

Lymphatic disorders in children can present significant clinical challenges, often requiring innovative and multidisciplinary approaches to treatment. Among these disorders, chylothorax and Kaposi form lymphangiomatosis (KLA) are particularly notable due to their complexity and the severity of their symptoms. Chylothorax, characterized by the accumulation of lymphatic fluid in the pleural cavity, can be either congenital or acquired and often leads to respiratory distress. Traditional management of this condition includes dietary modifications and drainage, but these measures are not always effective. Lymphatic duct embolization (LDE) offers a minimally invasive alternative with promising results.

Kaposi form lymphangiomatosis (KLA) is a rare and aggressive lymphatic anomaly that typically presents with nonspecific symptoms such as respiratory distress, thrombocytopenia, and pleural effusions. The complexity of KLA often necessitates multimodal treatment approaches. For instance, an 11-year-old girl with KLA and recurrent chylous pericardial effusions, refractory to medical therapy and pericardial drainage, was successfully treated with thoracic duct embolization and sclerotherapy, resulting in significant clinical improvement and a prolonged symptom-free period.

The methodologies employed in LDE have seen significant advancements, particularly in imaging techniques. Dynamic Contrast-Enhanced MR Lymphangiography (DCMRL) and Intranodal Lymphangiography are two such techniques that have significantly improved the diagnosis and treatment planning for lymphatic disorders. These imaging techniques allow for detailed visualization of the lymphatic system, aiding in the precise targeting of abnormal vessels during embolization procedures.

The embolization procedure itself typically involves accessing the thoracic duct or other lymphatic vessels under ultrasound and fluoroscopic guidance. Various embolic agents can be used in the procedure, including coils, n-butyl cyanoacrylate glue, and lipiodol. For instance, in a study involving neonatal chylothorax, lipiodol embolization demonstrated high effectiveness with minimal adverse effects. The success rates of LDE in treating pediatric

lymphatic disorders are promising. In one study, LDE achieved clinical success in 90% of patients with persistent chylothorax, highlighting its efficacy as a treatment option. Additionally, combining LDE with sclerotherapy has shown enhanced outcomes in cases of extensive lymphatic malformations.

The long-term prognosis for children undergoing LDE varies depending on the underlying condition and the extent of lymphatic involvement. For example, the aforementioned case of KLA treated with LDE, and sclerotherapy remained symptom-free for 15 months, suggesting that LDE can provide durable relief in some patients.

Recent advancements in therapeutic approaches have also contributed to the efficacy of LDE. Novel systemic therapies, such as mTOR inhibitors and MEK inhibitors, are being explored to complement LDE in treating lymphatic disorders. These therapies target the underlying molecular pathways involved in lymphatic anomalies, potentially improving outcomes when used in conjunction with embolization procedures.

The management of pediatric lymphatic disorders often requires a multidisciplinary approach, involving pediatricians, interventional radiologists, surgeons, and other specialists. Collaborative care ensures comprehensive treatment planning and follow-up, addressing both the immediate and long-term needs of the patient. This holistic approach is essential in managing the complex nature of lymphatic disorders in children.

The success rates of LDE in pediatric patients are particularly noteworthy. For instance, in a study of 88 patients with persistent chylothorax, LDE was clinically successful in 90% of the cases, underscoring its effectiveness. The procedure offers a valuable, minimally invasive treatment for a challenging and often morbid condition. Additionally, the combination of LDE with regional doxycycline sclerotherapy has shown enhanced outcomes, particularly in preventing recurrent chylopericardium by sclerosing the presumed nidus for lymphatic drainage.

The role of imaging in the success of LDE cannot be overstated. Techniques like Dynamic Contrast-Enhanced MR Lymphangiography (DCMRL) and Intranodal Lymphangiography have revolutionized the field by providing detailed images of the lymphatic system. These imaging modalities enable clinicians to pinpoint the exact location of lymphatic leaks and other abnormalities, facilitating targeted and effective treatment. For example, DCMRL involves the injection of gadolinium contrast into inguinal lymph nodes, allowing for clear visualization of lymphatic flow and potential obstructions.

Furthermore, the development of novel imaging techniques has paved the way for better classification and prognostication of lymphatic disorders. These advancements have led to a deeper understanding of the pathophysiological mechanisms underlying these conditions, thereby allowing for more precise and individualized treatment plans. For instance, isolated neonatal chylothorax, a condition characterized by respiratory distress due to pleural effusion, can now be effectively treated with oil-based contrast (lipiodol) embolization. This relatively simple and minimally invasive technique has transformed the treatment landscape for this condition, offering high success rates with minimal adverse events.

The integration of systemic therapies with LDE represents another significant advancement in the treatment of lymphatic disorders. mTOR inhibitors, such as sirolimus, have shown promising results in managing vascular and lymphatic anomalies. These inhibitors work by targeting the mammalian target of rapamycin (mTOR), a kinase involved in cell proliferation and angiogenesis. By inhibiting this pathway, mTOR inhibitors can reduce the volume of lymphatic tissue and alleviate symptoms. In a study involving patients with lymphatic anomalies, including KLA, sirolimus therapy resulted in significant clinical improvements and enhanced quality of life.

MEK inhibitors have also emerged as a potential therapeutic option for lymphatic disorders. These inhibitors target the MAPK/ERK pathway, which is involved in cell growth and differentiation. By inhibiting this pathway, MEK inhibitors can potentially reduce the progression of lymphatic anomalies and improve clinical outcomes. The integration of these systemic therapies with LDE offers a multifaceted approach to managing complex lymphatic disorders, addressing both the immediate symptoms and the underlying molecular mechanisms.

The importance of a multidisciplinary approach in managing pediatric lymphatic disorders cannot be overstated. The complexity of these conditions often requires the expertise of various specialists, including pediatricians, interventional radiologists, surgeons, and other healthcare professionals. Collaborative care ensures that all aspects of the patient's condition are addressed, from diagnosis and treatment planning to follow-up and long-term management. This comprehensive approach is crucial in optimizing outcomes and improving the quality of life for children with lymphatic disorders.

In conclusion, lymphatic duct embolization represents a significant advancement in the treatment of pediatric lymphatic disorders. Its minimally invasive nature, combined with high success rates and the potential for durable outcomes, makes it a valuable option for conditions like chylothorax and Kaposi form lymphangiomatosis. The integration of novel imaging techniques and therapeutic agents continues to enhance the efficacy and safety of LDE, promising a brighter future for pediatric patients with complex lymphatic anomalies. Ongoing research and multidisciplinary care are essential to further optimize these interventions and improve the quality of life for affected children.

References:

1- Itkin M: Interventional Treatment of Pulmonary Lymphatic Anomalies. *Tech Vasc Interv Radiol*, 19(4), 299-304, 2016

2- Itkin M: Magnetic Resonance Lymphangiography and Lymphatic Embolization in the Treatment of Pulmonary Complication of Lymphatic Malformation. *Semin Intervent Radiol*, 34(3), 294-300, 2017

3- Bundy JJ, Srinivasa RN, Gemmete JJ, Hage AN, Chick JFB: Vascular and lymphatic complications after thoracic duct cannulation. *J Vasc Surg Venous Lymphat Disord*, 6(6), 730-736, 2018

4- Majdalany BS, Saad WA, Chick JFB, Khaja MS, Cooper KJ, Srinivasa RN: Pediatric lymphangiography, thoracic duct embolization and thoracic duct disruption: a single-institution experience in 11 children with chylothorax. *Pediatr Radiol*, 48(2), 235-240, 2018

5- Kylat RI, Witte MH, Barber BJ, Dori Y, Ghishan FK: Resolution of Protein-Losing Enteropathy after Congenital Heart Disease Repair by Selective Lymphatic Embolization. *Pediatr Gastroenterol Hepatol Nutr*, 22(6), 594-600, 2019

6- Bundy JJ, Ootaki Y, McLean TW, Hays BS, Miller M, Downing T: Thoracic duct embolization in kaposiform lymphangiomatosis. *J Vasc Surg Venous Lymphat Disord*, 8(5), 864-868, 2020

7- Rabinowitz D, Dysart K, Itkin M. (2022). Neonatal lymphatic flow disorders: central lymphatic flow disorder and isolated chylothorax, diagnosis and treatment using novel lymphatic imaging and interventions technique. *Curr Opin Pediatr*, 34(2), 191-196, 2022
8- Laje P, Dori Y, Smith C, Pinto E, Taha D, Maeda K: Surgical Management of Central Lymphatic Conduction Disorders: A Review. *J Pediatr Surg*, 59(2), 281-289, 2024

Cardiac Tamponade

Cardiac tamponade is a critical condition characterized by the accumulation of fluid in the pericardial space, leading to increased intrapericardial pressure and restricted cardiac function. This review examines cardiac tamponade in children, focusing on its etiology, clinical presentation, diagnosis, and management, based on a synthesis of multiple studies and case reports.

Cardiac tamponade is a medical emergency that can be fatal without prompt intervention. It involves the accumulation of fluid, blood, or gas in the pericardial sac, which compresses the heart and impairs its ability to pump blood effectively. While often associated with adult conditions, cardiac tamponade can also occur in children, stemming from various etiologies including infections, trauma, malignancies, and iatrogenic causes.

The causes of cardiac tamponade in children are diverse and can be broadly categorized into infectious, neoplastic, traumatic, and iatrogenic origins. Infections leading to cardiac tamponade often include bacterial, viral, and fungal pathogens. For instance, bacterial infections such as Staphylococcus aureus, Streptococcus pneumoniae, and Mycobacterium tuberculosis have been implicated in pediatric cases.

Malignancies, particularly acute lymphoblastic leukemia, and other cancers are also significant contributors to pericardial effusion and subsequent tamponade in children. Traumatic causes, though less common, include blunt or penetrating chest injuries that result in hemopericardium. Iatrogenic causes are increasingly reported, especially related to central venous catheter placement, as evidenced by multiple case reports and studies.

The clinical presentation of cardiac tamponade in children can vary but typically includes symptoms such as dyspnea, tachycardia, and chest pain. Physical examination findings may reveal hypotension, jugular venous distension, distant heart sounds, and pulsus paradoxus (a decrease in systolic blood pressure during inspiration). In severe cases, signs of shock and hemodynamic instability are present.

A specific case involving a 13-year-old girl with multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19 presented with symptoms of myopericarditis and acute pericardial tamponade, highlighting the variability and complexity of clinical manifestations.

Diagnosis of cardiac tamponade in children involves a combination of clinical assessment and imaging techniques. Echocardiography is the gold standard for diagnosing pericardial effusion and tamponade, as it can visualize the fluid accumulation and assess its impact on cardiac function. Other imaging modalities such as chest X-ray and CT scans can provide additional information but are secondary to echocardiography.

Electrocardiography (ECG) often shows low voltage QRS complexes and electrical alternans (variation in QRS complex amplitude), which are suggestive of significant pericardial effusion and tamponade.

Management of cardiac tamponade in children requires prompt intervention to relieve the pressure on the heart. The primary treatment is pericardiocentesis, which involves the insertion of a needle into the pericardial space to aspirate the excess fluid. This procedure can be performed under echocardiographic guidance to increase its safety and efficacy.

In cases where pericardiocentesis is not feasible or unsuccessful, surgical options such as pericardial window or pericardiectomy may be necessary. These procedures create a continuous drainage route for the pericardial fluid, preventing reaccumulation.

Several case studies provide insights into the diverse presentations and management strategies for cardiac tamponade in children. For instance, a case involving a 7-year-old girl who developed tamponade following Hickman catheter insertion for bone marrow transplantation illustrates the risks associated with central venous catheter placement and the need for prompt surgical intervention.

Another report described a 9-year-old boy and an 11-year-old girl presenting with acute cardiac tamponade as the first manifestation of systemic lupus erythematosus (SLE). These cases underscore the importance of considering autoimmune disorders in the differential diagnosis of pericardial effusion in children.

Cardiac tamponade in children is a life-threatening condition that requires immediate recognition and intervention. The etiology is varied, encompassing infections, malignancies, trauma, and iatrogenic causes. Clinical presentation can be subtle, necessitating a high index of suspicion and prompt use of diagnostic imaging. Management primarily involves pericardiocentesis, with surgical options reserved for refractory cases. Continuous monitoring and follow-up are essential to prevent recurrence and manage underlying conditions. Further research and case documentation are needed to enhance understanding and improve outcomes for pediatric patients with cardiac tamponade.

References:

1- Fares, M.H., Itani, M.H., Malek, E.M., & Arabi, M.T: Cardiac tamponade as the first manifestation of systemic lupus erythematosus in children. BMJ Case Reports, doi:10.1136/bcr-2012-006927 , 2012
2- Singh, Y., Valverde, I., Sanchez-de-Toledo, J., et al: Acute cardiovascular manifestations in children with multisystem inflammatory syndrome associated with COVID-19 infection in Europe. Circulation, 143, 21-32, 2013
3- Valverde, I., Singh, Y., Sanchez-de-Toledo, J., et al: Acute cardiovascular manifestations in 286 children with multisystem inflammatory syndrome associated with COVID-19 infection in Europe. Circulation, 143, 21-32, 2016
4- Belhadjer, Z., Méot, M., Bajolle, F., et al: Acute heart failure in multisystem inflammatory syndrome in children in the context of Global SARS-CoV-2 pandemic. Circulation, 142, 429-436, 2020
5- Pawar, R.S., Wasgaonkar, G., Killedar, S., Bhoje, A., Patil, U: Multisystem Inflammatory Syndrome in Children Presenting as Cardiac Tamponade. The Pediatric Infectious Disease Journal, 40(12), e530-e531, 2021
6- Cevik, M., & Erek, E: Hickman catheter-induced cardiac tamponade-related cardiac perforation

management by mediastinotomy in children and a review of the literature. Trauma Case Reports, 32, 100436, 2021
7- Whittaker, E., Bamford, A., Kenny, J., et al: Clinical characteristics of children with a pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2. JAMA, 324, 259-269, 2021
8- Tomy, C., Hakeem, A.Y., Oladipo, O.O., et al: Pericardial diseases in COVID-19: a contemporary review. Current Cardiology Reports, 23, 90, 2024

Isolated Fallopian Tube Torsion

Isolated Fallopian Tube Torsion (IFTT) is an exceptionally rare gynecological emergency, particularly affecting adolescent females. The condition involves the twisting of the fallopian tube without involvement of the ovary, leading to acute abdominal pain and, if not treated promptly, can result in severe complications such as ischemia, necrosis, infertility, or infection. The incidence of IFTT is estimated at approximately 1 in 1,500,000, highlighting its rarity and the diagnostic challenges associated with it due to nonspecific symptoms. This review synthesizes findings from eight detailed reports spanning several years, offering a comprehensive understanding of IFTT's clinical presentation, diagnostic hurdles, management strategies, and outcomes.

Patients with IFTT typically present with acute lower abdominal pain, often accompanied by nausea and vomiting. The pain may be intermittent or constant, and it can radiate to the thigh or groin, exacerbating the diagnostic challenge. The nonspecific nature of these symptoms often leads to a wide differential diagnosis, including appendicitis, ovarian torsion, ruptured ovarian cysts, urinary tract infections, and gastrointestinal disorders. This overlap necessitates a high degree of clinical suspicion and often extensive diagnostic workup to identify IFTT accurately.

Imaging plays a crucial role in the diagnostic process, although it is often inconclusive. Ultrasound is usually the first-line imaging modality used to evaluate acute abdominal and pelvic pain. Specific ultrasound findings suggestive of IFTT include a tubular structure with thick echogenic walls, internal debris, and a narrowed end, known as the beak sign. Doppler ultrasound can also reveal reduced or absent blood flow in the torted tube, although these signs are not definitive. MRI and CT scans can provide additional detail but are often employed when ultrasound findings are ambiguous. Ultimately, diagnostic laparoscopy is frequently necessary to confirm the diagnosis, as it allows direct visualization of the twisted fallopian tube and excludes other potential causes of the symptoms.

The management of IFTT primarily involves surgical intervention. The preferred approach is laparoscopy, which is minimally invasive and allows for both diagnosis and treatment. During surgery, the torted tube is detorted, and any associated pathology, such as hydrosalpinx or paratubal cysts, is addressed. Hydrosalpinx, a condition where the fallopian tube is filled with serous fluid, is a common finding associated with IFTT and complicates its management. The decision to preserve or remove the fallopian tube depends on its viability post-detorsion and the presence of any irreversible damage. If the tube appears viable after detorsion, conservative management with tube preservation is preferred to maintain fertility.

However, if there are signs of necrosis or if the tube does not recover its normal appearance, salpingectomy (removal of the fallopian tube) is performed to prevent further complications.

The outcomes of IFTT management vary based on the timing of intervention and the condition of the fallopian tube at diagnosis. Early intervention generally results in better outcomes, with higher chances of tube preservation and subsequent fertility. Delayed diagnosis and treatment, on the other hand, often necessitate salpingectomy due to irreversible damage to the fallopian tube. Studies have shown that even when the fallopian tube appears ischemic, there can be significant recovery if detorsion is performed promptly. This underscores the importance of maintaining a high index of suspicion for IFTT in young females presenting with acute abdominal pain and pursuing early surgical evaluation.

The literature reveals that IFTT often occurs on the right side more frequently than the left. This could be due to the cushioning effect of the sigmoid colon on the left side, which potentially reduces the likelihood of torsion. Moreover, right-sided abdominal pain is more commonly investigated surgically due to the differential diagnosis that includes appendicitis, leading to more frequent identification of right-sided IFTT.

In pediatric populations, managing IFTT requires special consideration due to the implications for future fertility. Conservative surgical approaches are generally preferred to preserve the fallopian tube and maintain reproductive potential. However, in cases where hydrosalpinx is present, conservative management can be challenging. Hydrosalpinx increases the risk of recurrence of torsion, and patients often require multiple surgeries if the tube is initially preserved. Thus, a balance must be struck between preserving fertility and preventing recurrent torsion and associated complications.

Case studies and retrospective reviews have highlighted the varied presentations and management outcomes of IFTT. In a series of cases, patients ranged from 6 to 16 years old, with most presenting with acute right lower quadrant pain and some exhibiting additional symptoms like thigh pain or dysuria. Diagnostic imaging often revealed adnexal masses or hydrosalpinx, leading to surgical exploration. The intraoperative findings confirmed isolated fallopian tube torsion in all cases, with varied management strategies based on the viability of the fallopian tube and associated pathologies.

One notable case involved a 15-year-old girl who presented with four days of intermittent abdominal pain exacerbated by movement. Imaging revealed bilateral paratubal cysts, and diagnostic laparoscopy confirmed right IFTT. The right fallopian tube was detorted and preserved, and the paratubal cyst was enucleated. Follow-up showed no recurrence of symptoms, illustrating the potential for successful conservative management in select cases.

Another case involved an 11-year-old girl with acute right lower quadrant pain and emesis. Imaging suggested bilateral paraovarian cysts, and surgery revealed right fallopian tube torsion with ischemia. Despite detorsion, the tube was non-viable and was resected. This

case underscores the need for prompt surgical intervention to assess and manage the viability of the fallopian tube.

In conclusion, Isolated Fallopian Tube Torsion is a rare but significant cause of acute abdominal pain in adolescent females, with important implications for fertility if not promptly diagnosed and treated. High clinical suspicion, appropriate imaging, and timely surgical intervention are critical to managing this condition effectively. While conservative management with tube preservation is ideal, especially in pediatric patients, the presence of associated conditions like hydrosalpinx may necessitate more aggressive surgical approaches to prevent recurrence and ensure patient well-being. Future research should focus on refining diagnostic criteria and management protocols to improve outcomes for patients with this rare condition.

References:

1- Bertozzi, M., Magrini, E., Riccioni, S., Giovannali, P., & Appignani, A: Isolated fallopian tube torsion with hydrosalpinx: Review of a debated management in a pediatric population. *Journal of Pediatric Surgery*, 52(10), 1553-1560, 2017

2- Spinelli, C., Piscioneri, J., & Strambi, S: Adnexal torsion in adolescents: update and review of the literature. *Current Opinion in Obstetrics and Gynecology*, 27(5), 320-325, 2018

3- Raban, O., Zilber, H., Hadar, E., Efrat, Z., Krissi, H., Wiznitzer, A., Meizner, I., & Bardin, R: Isolated Fallopian Tube Torsion: A Unique Ultrasound Identity or a Serial Copycat? *Journal of Ultrasound in Medicine*, 37(10), 2387-2393, 2018

4- Bertozzi, M., Noviello, C., Molinaro, F., Ratta, A., Lisi, G., Cobellis, G., Federici, S., Chiesa, P.L., Martino, A., & Messina, M: Isolated fallopian tube torsion in pediatric age: An Italian multicenter retrospective study. *Journal of Pediatric Surgery*, 55(4), 711-714, 2020

5- Baracy Jr, M.G., Hu, J., Ouillette, H., & Aslam, M.F: Diagnostic dilemma of isolated fallopian tube torsion. *BMJ Case Reports*, 14(7), e242682, 2021

6- Crosier, C.J., & Ricca, R.L: Isolated Fallopian Torsion: A Pediatric Case Series. *The American Surgeon*, 89(9), 3917-3919, 2023

7- Mariani, A., Hameury, F., Dubois, R., Demède, D., Gelas, T., Mure, P.Y., & Gorduza, D: Isolated Fallopian Tube Torsion in Children with Hydrosalpinx: Is Conservative Management an Option? *Pediatric Emergency Care*, doi: 10.1097/PEC.0000000000003209, 2024

8- Schwartz, B., Weerasooriya, N., Mercier, R., Gould, S., Saul, D., & Berman, L: Factors Associated With Isolated Fallopian Tube Torsion in Pediatric Patients. *Journal of Pediatric Surgery*, 59(8), 1538-1544, 2024

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