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Managing Idiopathic Retroperitoneal Fibrosis: A Multidisciplinary Approach

Learning Objective: At the conclusion of this continuing medical education activity, the participant will be able to initiate an appropriate workup of retroperitoneal fibrosis (RPF), distinguish idiopathic RPF from other causes of RPF, and develop a medical and surgical treatment plan for patients with idiopathic RPF.

This AUA Update aligns with the American Board of Urology Module on Calculus, Laparoscopy-Robotics and Upper Tract Obstruction. Additional information on this topic can be found in the AUA Core Curriculum sections on Laparoscopy and Robotic Surgery, and Reconstructive Urology.

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KEY WORDS: retroperitoneal fibrosis, autoimmune, ureteral obstruction, ureterolysis

This article is dedicated to the memory of the late nephrologist, Rick Swartz, MD, whose invaluable expertise in idiopathic retroperitoneal fibrosis, dedication to care of this complex patient population, and collaborative spirit greatly impacted both patients and collaborators at the University of Michigan and around the world.

DEFINITION AND ETIOLOGY

Idiopathic retroperitoneal fibrosis (iRPF) is an autoimmune retroperitoneal fibro-inflammatory disease distinct from other identifiable causes of retroperitoneal fibrosis (RPF). Diagnoses that may masquerade as iRPF include hematological or metastatic malignancy, sarcoma, granulomatosis, or vasculitis, or RPF related to prior surgical, radiation-based, or chemotherapy treatments. A clear identifiable cause is found in less than 25%-30% of RPF cases, and it is therefore categorized as idiopathic with multiple proposed mechanisms such as occult autoimmune disease or vasculitis, local immunological reaction to arterial atherosclerosis, and others.[1] Initial unpublished descriptions of the disease were made as early as 1905 by Joaquin Albarran, a Cuban-born urologist practicing in France. In 1948, John Ormond published a case report while at Henry Ford Hospital further describing RPF and detailing suspicion that ureteral obstruction was secondary to retroperitoneal inflammation, earning him the "Ormond's disease" eponym.[2]

iRPF is uncommon, with an estimated incidence of 1:100,000. A majority of cases involve periaortic inflammatory changes at the level of the infrarenal abdominal aorta, with variable histopathological findings including nonspecific inflammation, histiocytic cells, and granuloma. Previously identified risk factors include smoking,[3] with a reported OR of 3.15 for exposure to cigarettes >5 years.[4] Asbestos is also associated with iRPF, and the combination of both asbestos and tobacco exposure conferred an OR of 13.5.[4] Males predominate in iRPF.[5]

DIAGNOSIS AND WORKUP

A typical presentation involves constitutional symptoms such as weight loss, fatigue, and abdominal or back pain. Ureteral involvement and consequent obstruction can occur in upwards of 80% of cases, in which case hydronephrosis or renal dysfunction can be discovered on workup of these symptoms. Scrotal pain, hydrocele, and varicocele have also been reported due to gonadal vein involvement by the retroperitoneal mass.[5] Additional presenting symptoms can include sequelae of gastrointestinal, biliary, or pancreatic involvement, or lower extremity venous or arterial compression.

Laboratory assessment with inflammatory markers, though nonspecific, is useful for purposes of monitoring response to medical treatment and relapse. Erythrocyte sedimentation rate and C-reactive protein are typically employed and should be obtained when iRPF is suspected, along with consideration of IgG1-4 and C3. IgG4-positive cases are an interesting subtype of iRPF, which account for a minority of overall cases, with 20%-56% of patients with IgG4-related disease demonstrating retroperitoneal involvement.[6] Other manifestations of this disease include inflammatory pseudotumors, hepatitis, and pancreatitis.[7] IgG4-positive vs -negative disease was compared in a 2021 study by Li et al with findings of more severe fibrosis in IgG4+ patients, higher recurrence rate, higher incidence of renal failure, and male predominance.[8] In such patients, IgG4 can serve as a helpful biomarker for disease progression or response to treatment. This population may be well served by tailored medical management with medications such as rituximab, an anti-CD20+ monoclonal antibody that is utilized in other related autoimmune disorders.

CT with IV contrast or MRI with gadolinium-based contrast is the preferred imaging modality for accurate initial assessment of RPF. In the event of acute or chronic renal dysfunction and inability to perform MRI, noncontrast CT is an acceptable alternative. **Cross-sectional imaging findings can differentiate iRPF from retroperitoneal malignancy. Characteristic findings in iRPF include uniform thickness around the aorta, increased cranio-caudal length of fibrosis, fibrosis at the level of the common and external iliac vessels, medial ureteral deviation, and hydronephrosis. In a study by Cohan et al, 50% of patients with iRPF or with IgG4-positive disease had such classic appearances compared to 8% of cases with biopsy proven malignancy.**[9] Further, presence of a circumferential aortic mass without vessel elevation was present in only 2% of malignancy cases. Atypical findings concerning for malignancy include regional lymphadenopathy, mass effect, or abnormal location of the retroperitoneal mass. Renal atrophy may be noted at time of diagnosis and has been shown to be present in 35% of cases.[10] An assessment of renal function with nuclear renography should also be obtained to determine the utility of renal preservation of the affected kidney.

Retroperitoneal mass biopsy can be performed by CT guidance, laparoscopically or robotically, or open, which serves primarily to rule out malignancy. **However, in the presence of "typical" iRPF imaging findings, biopsy can be safely avoided and reserved for cases of radiological uncertainty.** This was demonstrated by Cohan et al in a blinded study of suspected RPF cases wherein 3-month follow-up imaging was sufficient to rule out evolving malignancy.[9] When biopsy is performed, aside from a possible finding of IgG4-related disease, histopathology is nonspecific in iRPF and unhelpful in therapeutic decision-making.

TREATMENT: ACUTE AND LONG-TERM

Involvement of 1 or both ureters occurs in up to 70%-80% of iRPF cases. Consequent obstruction and acute kidney injury or failure may necessitate renal decompression with either ureteral stent or nephrostomy tube placement. Once decompression occurs, the focus of management becomes clarifying the diagnosis if still uncertain, removing offending pharmacological

ABBREVIATIONS: idiopathic retroperitoneal fibrosis (iRPF), mycophenolate mofetil (MMF), retroperitoneal fibrosis (RPF)

agents if present (eg, methysergide, pergolide), and addressing the retroperitoneal disease.

The role of medical management is to control the fibrosis and inflammation, reduce the retroperitoneal mass burden, and facilitate stent or nephrostomy tube removal in cases of ureteral obstruction by pharmacological means. A standard approach to medical management starts with a short-term corticosteroid course for 3-6 months, which typically results in relief of pain within several weeks. Our institutional protocol involves starting prednisone 30-60 mg daily for 2-4 weeks, with subsequent taper by 50% every 2 weeks until down to 10 mg daily for up to 3 months. However, corticosteroids alone are often insufficient to resolve ureteral obstruction.[11] Corticosteroid monotherapy typically requires a prolonged course, as reported by Al-Hammouri et al,[12] with an extended 24-month regimen, which puts the patient at risk for steroid-related toxicities.[13-15] A modern approach involves addition of steroid-sparing agents to further arrest the autoimmune disease, and to taper and discontinue steroids as early as possible to minimize treatment toxicity.[10] A typical treatment duration is 1-3 years depending on patient response or surgical treatment. There are multiple nonsteroidal agents that have been studied in this space. Mycophenolate mofetil (MMF) is commonly used and has well-documented tolerability and safety profiles within the transplant literature.[16-18] MMF inhibits T- and B-cell proliferation and is typically dosed at 1,000 mg twice daily. Other options include methotrexate, azathioprine, and cyclophosphamide, though the body of evidence supporting their use is limited in comparison to MMF. Agents such as tamoxifen or colchicine are less favored given absence of immunosuppressive action. Efficacy with tamoxifen or colchicine has only been observed in conjunctive use with corticosteroids, and the only randomized controlled trial of iRPF treatment strongly suggested superiority of corticosteroids when compared with tamoxifen.[19] As previously stated, rituximab can be considered in situations of IgG4-related disease or concomitant rheumatological diseases such as antineutrophil cytoplasmic antibody-associated vasculitis or rheumatoid arthritis.

MEDICAL MANAGEMENT OUTCOMES

Definitions of success and of relapse of medical management of iRPF vary in the literature, including radiographic changes related to retroperitoneal mass size, changes in hydronephrosis, inflammatory marker trends, and renal function changes. When looking specifically at patients with ureteral obstruction, we defined this as resolution of ureteral obstruction permitting stent removal without relapse in those initially stented.[10] Relapse was defined as recurrence of obstruction requiring ureteral decompression with ureteral stent or nephrostomy tube. Other relevant morbid events to consider when managing iRPF patients include medication side effects, nephrostomy tube placement for stent failure, and intolerance to stent or nephrostomy tube with desire for removal.

Medical management outcomes are best discussed separately by agent, and we will focus this discussion on outcomes related to ureteral obstruction for relevance to the urology audience. With regard to MMF, there are 3 series which give insight to the efficacy of this agent in iRPF. **Utilizing the previously mentioned definition of success, our 2021 series in 50 patients (87 ureters, 86% treated with MMF) demonstrated success in 69% with mean stent duration of 16 months.[10] Stent failure requiring nephrostomy tube occurred in 6% of patients. Relapse occurred in 18% of patients at a median of 3.8 years, and ultimately 8 of those patients underwent ureterolysis, of whom 3 experienced relapse or persistent obstruction postoperatively. Medication side effects requiring discontinuation occurred in 12% of patients.** Adler et al found 71% success in 7 patients with bilateral ureteral obstruction with a mean stent duration of 5.5 months, and Scheel et al saw 86% of ureters stent-free in 16 patients after a mean stent duration of 13.4 months.[17,18]

Utilizing corticosteroids alone for an extended 2-year period, Al-Hammouri et al demonstrated stent removal in 86% of obstructed ureters with unspecified stent duration,[12] and in a corticosteroid-predominant series with shorter treatment duration, Moriconi et al showed a 55% success rate with relapse in 39% of patients.[20] In a mixed regimen cohort (tamoxifen, azathioprine, MMF), Raffiota et al showed a 72% success rate with relapse in 38%.[21] A comparison of series is shown in the Table.

Ultimately, when limiting corticosteroid toxicity to patients, an MMF-based treatment regimen has demonstrated durability and efficacy in controlling iRPF and facilitating ureteral stent or nephrostomy tube removal.

FOLLOW-UP AND SURVEILLANCE

Retrograde pyelography at time of ureteral stent exchanges and interval cross-sectional imaging provide the best insight to changes in ureteral involvement during medical management. Retrograde pyelography involves a subjective assessment of ureteral compression, upstream dilation, and drainage of instilled contrast. Presently, there are no validated intraoperative assessments to guide clinical decision-making of whether or not stent removal is appropriate. Our clinical practice is to instill 10-15 cc diluted contrast retrograde via a ureteral catheter placed distal to the ureteral involvement. If contrast passes up to the proximal collecting system, the ureter is fluoroscopically observed periodically over 2 minutes. If a substantial amount of antegrade passage of contrast is noted, the ureter is likely ready for stent removal. Surveillance with ultrasound and renal function labs are performed 1-2 weeks after stent removal to ensure ureteral patency. As previously mentioned, inflammatory markers may also be helpful in deciding upon stent removal timing. In our series, we found 53%, 65%, and 47% decreases from peak levels in erythrocyte sedimentation rate, C-reactive protein, and IgG4, respectively, during the course of medical treatment, with coincidental low levels for each within 3 months of stent removal. While not our standard institutional practice, in cases of persistent obstruction where reconstructive surgery is not feasible and long-term stent exchange is planned, metallic ureteral stents can be considered to facilitate longer periods between exchanges.[22] In situations of persistent hydronephrosis or persistent renal dysfunction compared to baseline function with a ureteral stent in place, nephrostomy tube placement can be considered for maximal decompression and recovery of renal function, particularly to ensure all pharmacological options are available for medical management (Figure 1).

Table. Medical Management Outcomes

	No. patients with ureteral stent for UO	Successful stent removal with medical management (%)	Stent duration (mo)[a]	Relapse (%)	Time to relapse (y)	Ureterolysis	Nephrectomy (%)
Santiago et al[10]	50 (87 ureters)	68	16	18	3.8	15%	7.6
Raffiota et al[21]	22	72	–	38	5.19	5/35 Original cohort	–
Moriconi et al[20]	20	55	–	39	1.25	3/43 Original cohort	–
Al-Hammouri et al[12]	92 (160 ureters)	N/A 82.5% of obstructed ureters stent-free	–	17	–	–	–
Scheel et al[17]	16	N/A 86% of obstructed ureters stent-free	13.4 (mean)	–	–	–	–
Adler et al[18]	7	71	5.5 (mean)	–	–	–	–

Abbreviations: N/A, not available; UO, ureteral obstruction.
[a]Stent duration specifically in patients successfully managed medically.

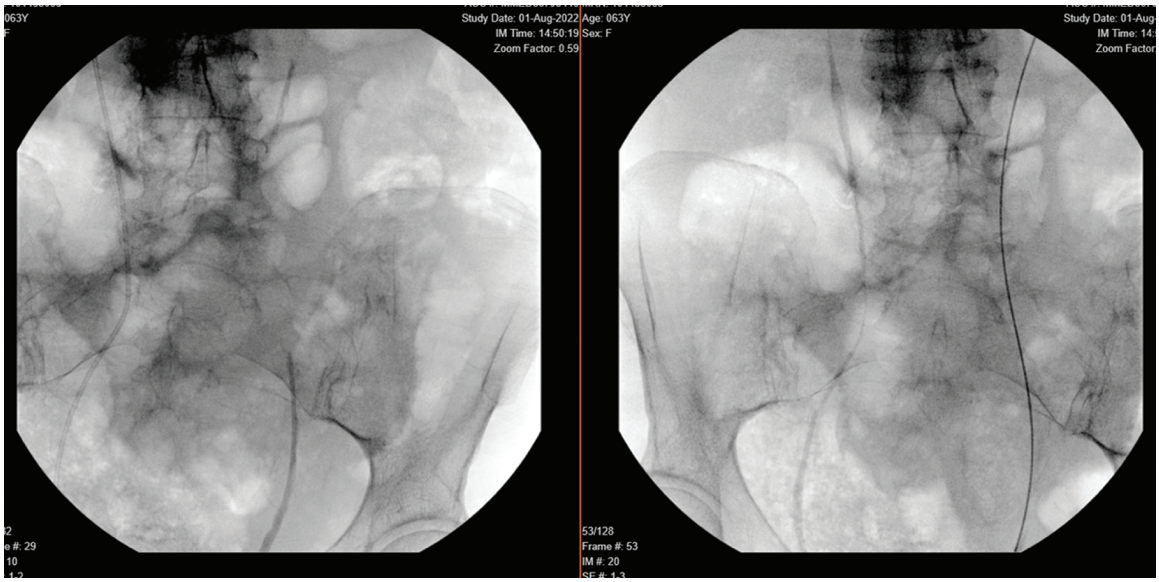


Figure 1. Retrograde pyelography in a patient on mycophenolate mofetil for 1 year. Resolved right ureteral obstruction (right stent removed, right panel) and persistent left obstruction (left stent exchanged, left panel).

SURGICAL TREATMENT

Prior to the advent of medical treatment utilization, ureterolysis was the mainstay treatment for iRPF. Although ureterolysis will address the obstruction temporarily, the inflammatory disease process persists without medical treatment. Despite this, ureterolysis remains a valuable treatment option and for some patients is the solution to renal decompression–related complications. Surgical management beyond renal decompression such as ureterolysis or nephrectomy should be particularly considered in patients fit for surgery in several cases: prolonged need for ureteral stent or nephrostomy tube beyond 2 years of continued medical management; poor tolerance of medications; demonstrated atrophic, nonfunctioning kidney with or without symptoms or infection; and stent or nephrostomy tube symptoms significantly impacting quality of life. Nephrectomy was performed in 7.6% of patients from our cohort,[10] and in addition to cases of atrophic kidney, can also be considered in cases of nonreconstructable ureter. Of note, our practice is to remove the stent at least 3-4 weeks preoperatively to reduce periureteral inflammatory rind.

Ureterolysis involves ureteral identification and dissection of the ureter from the retroperitoneal mass until freely mobile, extending caudally and cranially until healthy periureteral tissue is reached. Commonly an omental wrap with ureteral lateralization or alternatively intraperitonealization is employed to distance the ureter from the fibrotic mass,

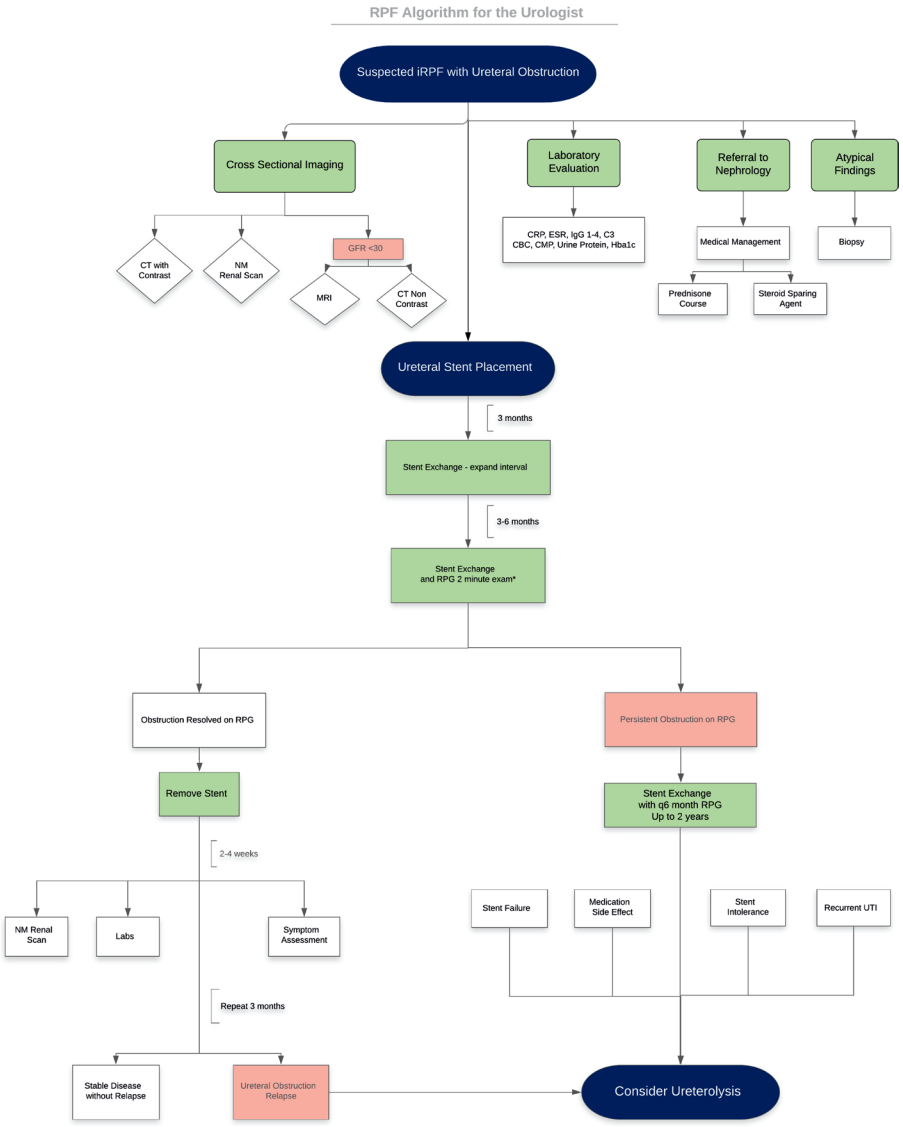
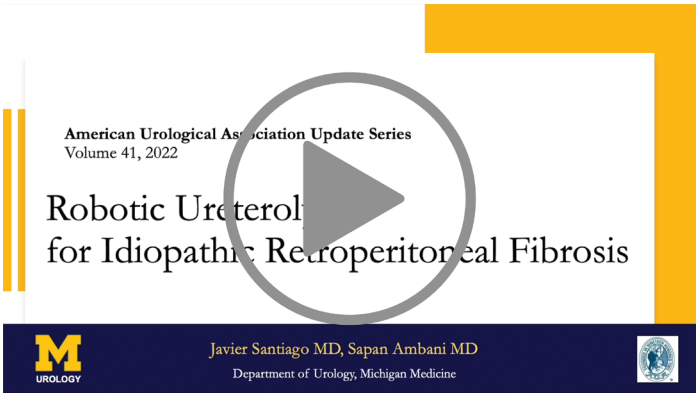


Figure 2. Management algorithm. CBC indicates complete blood count; CMP, comprehensive metabolic panel; CRP, C-reactive protein; CT, computerized tomography; ESR, erythrocyte sedimentation rate; GFR, glomerular filtration rate; iRPF, idiopathic retroperitoneal fibrosis; MRI, magnetic resonance imaging; NM, nuclear medicine; q, every; RPF, retroperitoneal fibrosis; UTI, urinary tract infection.

as described by Miles et al in 1984[23] and reemphasized by Lindell and Lehtonen in 1998.[24] While iRPF is a rare entity, several modern series exist in the literature for open, laparoscopic, and robotic approaches. Worth noting is that some cases with significant ureteral stricture may necessitate additional reconstructive techniques such as reimplantation with a Boari flap or psoas hitch, ureteroureterostomy, buccal ureteroplasty, and others. Patients should be counseled on the potential need for such procedures to reestablish ureteral patency.

The largest open series from O'Brien and Fernando reports 50 patients who underwent open ureterolysis after initial ureteral stent placement and corticosteroid monotherapy with 12-month follow-up.[25] They demonstrated 94% stent-free status at 1 year along with durable improvements in renal function. With longer follow-up of 25 months, Zahran et al demonstrated 78% stent-free status after open ureterolysis with omental wrap in 21 patients with a 6- to 12-month corticosteroid course.[26] Superior renal function was seen in the ureterolysis group when compared to corticosteroid monotherapy with ureteral stent. Srinivasan et al compared open to laparoscopic technique in a series comprised of iRPF in 50% of cases, finding that in the iRPF subgroup (8 open, 27 laparoscopic) there was lower incidence of blood transfusion and shorter length of stay with a laparoscopic approach.[27] Resolution of ureteral obstruction was reported as 100%, although with unclear follow-up, relapse, or postoperative pharmacological management. Additional small series include those of Fugita[28] and Simone[29] et al, which report 75% and 100% successful ureterolysis in iRPF patients, respectively. Both studies also confirm the appropriateness of omitting contralateral ureterolysis in an unaffected ureter. Comparing studies using open and laparoscopic technique, estimated blood loss and length of stay were 300-390 vs 75-300 cc and 4-8 vs 3.3-5.5 days, respectively.[24,26-28]

Regarding a robotic approach, some relevant advantages include precise dissection of scarred tissue planes–particularly in perivascular spaces–as well as usage of fluorescent dyes such as indocyanine green or fluorescein to aid in identification and dissection of the ureter. Comparable outcomes are suggested, with 2 robotic series reporting 70%-97% resolution of ureteral obstruction (heterogenous RPF groups), and estimated blood loss and length of stay of 57-86 cc and 2.6-3.1



Video. Robotic ureterolysis for idiopathic retroperitoneal fibrosis. Video available at https://auau.auanet.org/US2023-L24.

days (Keehn[30] and Marien[31] et al). **In comparison, a robotic approach, when available and within the tool kit of the operating surgeon, appears to confer lower blood loss, shorter length of stay, and less overall morbidity without compromising success.**

CONCLUDING THOUGHTS AND APPROACH

When considering the available literature on ureterolysis in iRPF, conclusions are limited by length of follow-up and inconsistency in approach to, and variable use of, medical treatment. Our institutional approach is to provide early anti-inflammatory and immunomodulation therapy in order to control the disease process and potentially resolve ureteral involvement and obstruction without needing to proceed with ureterolysis, which has the capacity to be a technically challenging and potentially morbid operation. We suggest a multidisciplinary approach to cases of iRPF with ureteral involvement, with joint workup and comanagement by urology, nephrology, and/or rheumatology that involves the following principles:

- Careful initial scrutiny for atypical clinical and radiographic features that warrant biopsy
- Serological evaluation for underlying immunological disease
- Renal decompression for ureteral obstruction and renal dysfunction
- Assessment for renal atrophy and function of affected renal unit
- Initial corticosteroid treatment with early employment of a steroid-sparing agent and tapering of said steroids
- Follow-up imaging within 3-6 months of initial presentation to confirm absence of atypical radiographic features such as retroperitoneal mass growth concerning for underlying malignancy
- Monthly or bimonthly assessment of inflammatory markers to monitor response to treatment
- Reassessment of ureteral obstruction at planned 3- to 6-month intervals with retrograde pyelography and ureteral stent exchange vs removal
- Ureterolysis/ureteral reconstruction or nephrectomy for persistent ureteral obstruction beyond 1-2 years despite medical treatment or in patients who do not tolerate ureteral stents or nephrostomy tubes (Figure 2 and Video).

DID YOU KNOW?

- Idiopathic retroperitoneal fibrosis is an uncommon autoimmune disease which commonly leads to ureteral obstruction and can be accurately diagnosed clinically and radiographically in a majority of cases.
- Medical management with short-term corticosteroids followed by an extended course of a steroid-sparing agent can reduce retroperitoneal mass burden and resolve ureteral obstruction in over 50% of patients.
- When medical management is ineffective or not tolerated, ureterolysis is a very effective surgery to resolve ureteral obstruction vs nephrectomy in the setting of atrophic kidney.

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Study Questions Volume 42 Lesson 24

1. On cross-sectional imaging, which of the following findings is most suggestive of idiopathic retroperitoneal fibrosis?
   a. Regional lymphadenopathy
   b. Medialization of the ureter
   c. Mass effect from retroperitoneal mass
   d. Asymmetrical retroperitoneal mass

2. Which of the following is a risk factor for iRPF?
   a. Female gender
   b. Alcohol abuse
   c. History of chemotherapy
   d. Smoking

3. During evaluation, which of the following are the recommended iRPF-specific laboratory studies for workup?
   a. Erythrocyte sedimentation rate, C-reactive protein, IgG1-4
   b. Alpha-fetoprotein, beta-human chorionic gonadotropin
   c. Antinuclear antibody, p-antineutrophil cytoplasmic antibody
   d. Lactate dehydrogenase

4. In the management of iRPF, what percentage of patients had resolution of ureteral obstruction with medical management alone?
   a. 25%
   b. 40%
   c. 69%
   d. 90%

5. In the surgical management of iRPF, what technique is recommended at the time of ureterolysis?
   a. Intraperitonealization
   b. Omental wrapping of the ureter
   c. Nephropexy
   d. Regional lymphadenectomy