Emphysematous pyelonephritis

PRESENTER
Dr. Ershad Hussain

2nd year Post Graduate,
Department of Urology

Narayana Medical College

MODERATOR
Dr.Vedamurthy Reddy

Professor and HOD, Dept Of Urology

Narayana Medical College

Introduction

Emphysematous pyelonephritis (EPN) is an acute severe necrotizing infection of the renal parenchyma and its surrounding tissues that results in the presence of gas in the renal parenchyma, collecting system or perinephric tissue.

The first case was reported by Kelly and MacCullum in 1898.

Since then terms such as ‘renal emphysema’, ‘pneumonephritis’ as well as ‘emphysematous pyelonephritis’ have been used to describe the gas-forming infection.

In 1962, Schultz and Klorfein suggested the use of ‘emphysematous pyelonephritis’ as the preferred term, as it emphasizes the relationship between infective pathology and gas formation.

Gas in the collecting systems only, ‘emphysematous pyelitis’, is a separate condition and could be secondary to instrumentation of the urinary tract.

A patient infected with emphysematous pyelitis has an excellent prognosis with medical management (MM).

Whereas EPN deserves special attention because of its life threatening potential with requirement of either MM or surgical management immediately.

Mortality from EPN is primarily attributable to septic complications.

EPN was associated with a mortality rate of up to 78% until the late 1970s but, over the last two decades, improvement in management techniques has reduced the mortality rate to 20-25%

Associated factors

Diabetes mellitus is the single most common associated factor.

Up to 95% of patients with EPN have underlying uncontrolled diabetes mellitus.

Other reported factors associated with the development of EPN are

- Drug abuse,
- Neurogenic bladder,
- Alcoholism,
- Anatomic anomaly like polycystic kidney disease

There is a preponderance of EPN in females; the female:male ratio reported in various studies is 6 : 1.

Increased susceptibility to UTI seems to be the reason for the higher incidence in females.

The risk of developing EPN secondary to a urinary tract obstruction is ≈ 25–40%.

Although cases of EPN have been reported worldwide, it appears to be geographically more common in Asia with most case studies being reported from there.

The left kidney was more frequently involved than the right one (60-70%), Less than 10% of cases involve both kidneys.

AETIOLOGY & PATHOGENESIS

Emphysematous pyelonephritis is a severe, necrotizing form of acute bacterial pyelonephritis and *Escherichia coli* remains the most common causative pathogen; the organism has been isolated on urine or pus cultures in nearly 70% of the reported cases.

There have, however, been reports of *Proteus mirabilis, Klebsiella pneumoniae*, Group D *Steptococcus* and coagulase-negative *Staphylococcus* being the causative agent for EPN.

Anaerobic microorganisms including *Clostridium septicum, Candida albicans, Cryptococcus neoformans* and *Pneumocystis jiroveci*, in rare cases, have been reported as the causative pathogen for EPN

Various factors involved in the pathogenesis of this condition have been suggested

- High levels of glucose within the tissues,
- The presence of gas-forming microorganisms,
- Impaired vascular blood supply,
- Reduced host immunity,
- The presence of obstruction within the urinary tract.

A high level of tissue glucose in association with impaired blood supply to the kidneys, which is prevalent in patients with diabetes, facilitates the process of anaerobic metabolism.

Gram-negative facultative anaerobic microorganisms such as *E. coli* are responsible for the production of gas via the fermentation of glucose and lactate.

This process results in the production of high levels of carbon dioxide and hydrogen which accumulate at the site of inflammation.

Radiologically guided needle aspiration of the gases released by the tissues was analysed by Huang and Tseng who found carbon dioxide and hydrogen to be the main constituents.

Nitrogen and oxygen have also been found along with traces of ammonia, methane and carbon monoxide.

Gas may extend beyond the site of inflammation to the sub-capsular, perinephric and pararenal spaces.

In some cases, gas was found to be extending into the scrotal sac and spermatic cord due to dissemination of infecting organisms.

In addition, the hypoxic environment of kidney medulla, especially in diabetic nephropathy with associated microvascular disease, is thought to predispose these patients to tissue ischemia and necrosis, thus potentiating the growth of gas-forming microbes.

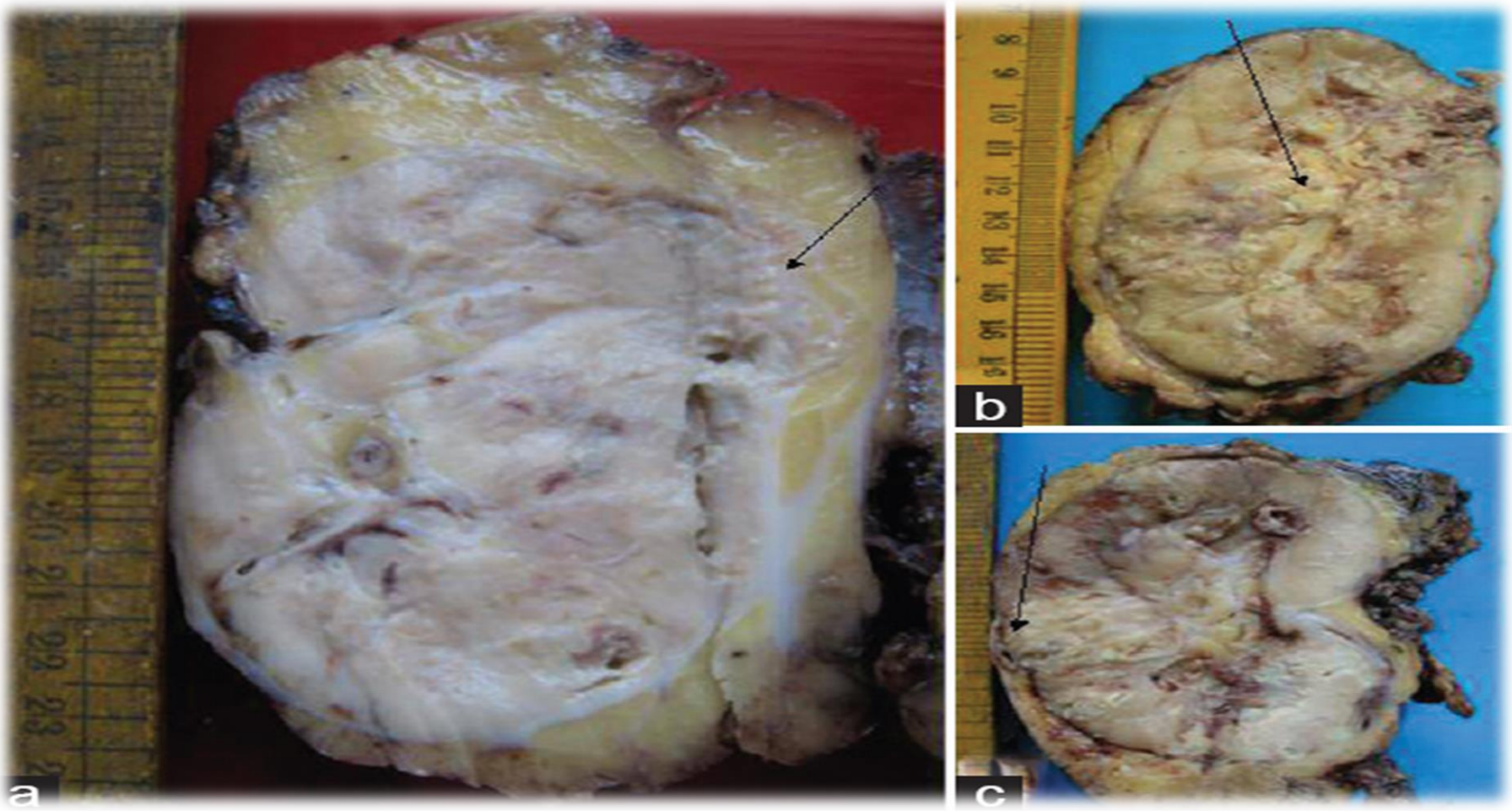
In addition to diabetes, many patients have urinary tract obstruction associated with urinary calculi or papillary necrosis and significant renal functional impairment.

EPN is also seen in patients of end-stage renal disease and immuno-compromised patients.

Pathological examination of the kidney reveals features of

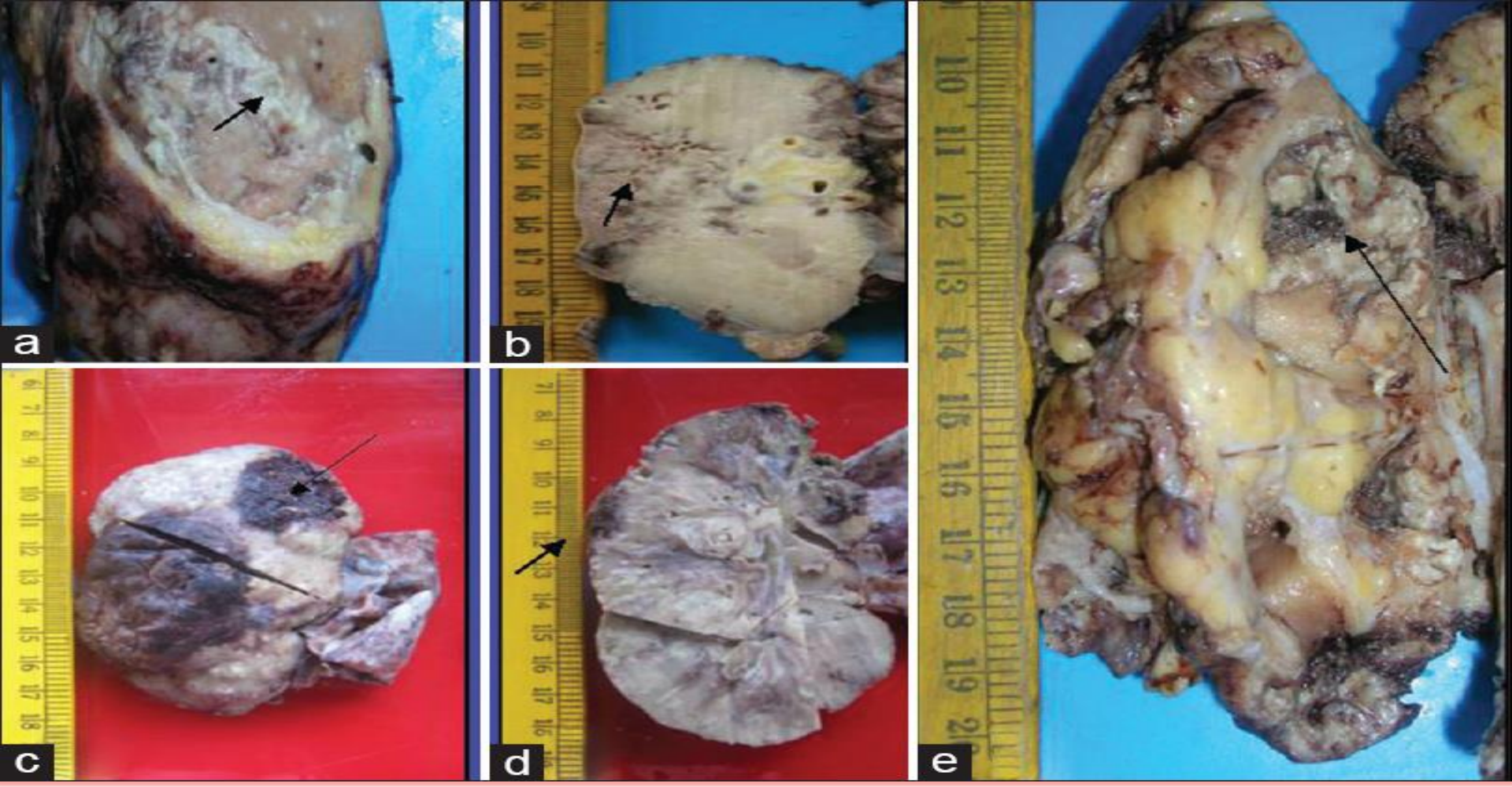
- abscess formation,
- foci of infarctions,
- vascular thrombosis,
- numerous gas-filled spaces and
- areas of necrosis surrounded by acute and chronic inflammatory cells implying septic infarction.

H&E stains reveal extensive suppurative inflammation, necrosis, abscess formation and gas bubbles. Large colonies of Gram negative bacilli can also be seen.



Gross examination of emphysematous pyelonephritis:

- specimen of enlarged kidney with loss of cortex and medulla with adherent perirenal fatty tissue (highlighted by arrow),
- foci of abscess within parenchyma (highlighted by arrow),
- abscess extending toward capsule (highlighted by arrow)



(a) confluent abscess seen on external surface of the kidney,

(b) friable necrotic parenchyma (arrow),

(c and d) hemorrhagic infarct on external surface and cut surface of the kidney,

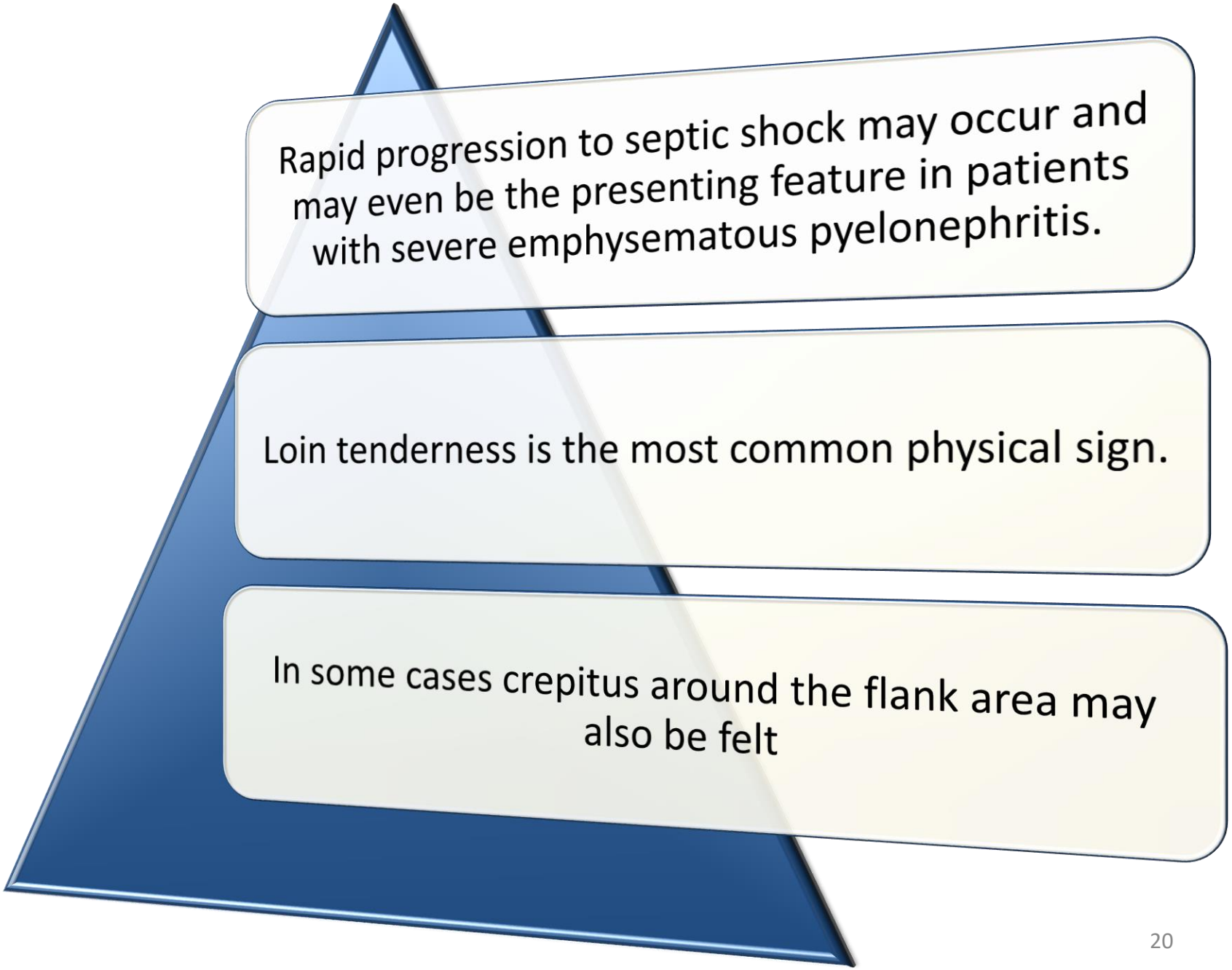
(e) a pyonephrotic cavity with purulent exudate lining the cavity

Presentation

Most patients present in the fourth or fifth decade.

The presenting physical symptoms are those of pyelonephritis such as dysuria, fever/rigours, nausea, vomiting, and flank pain.

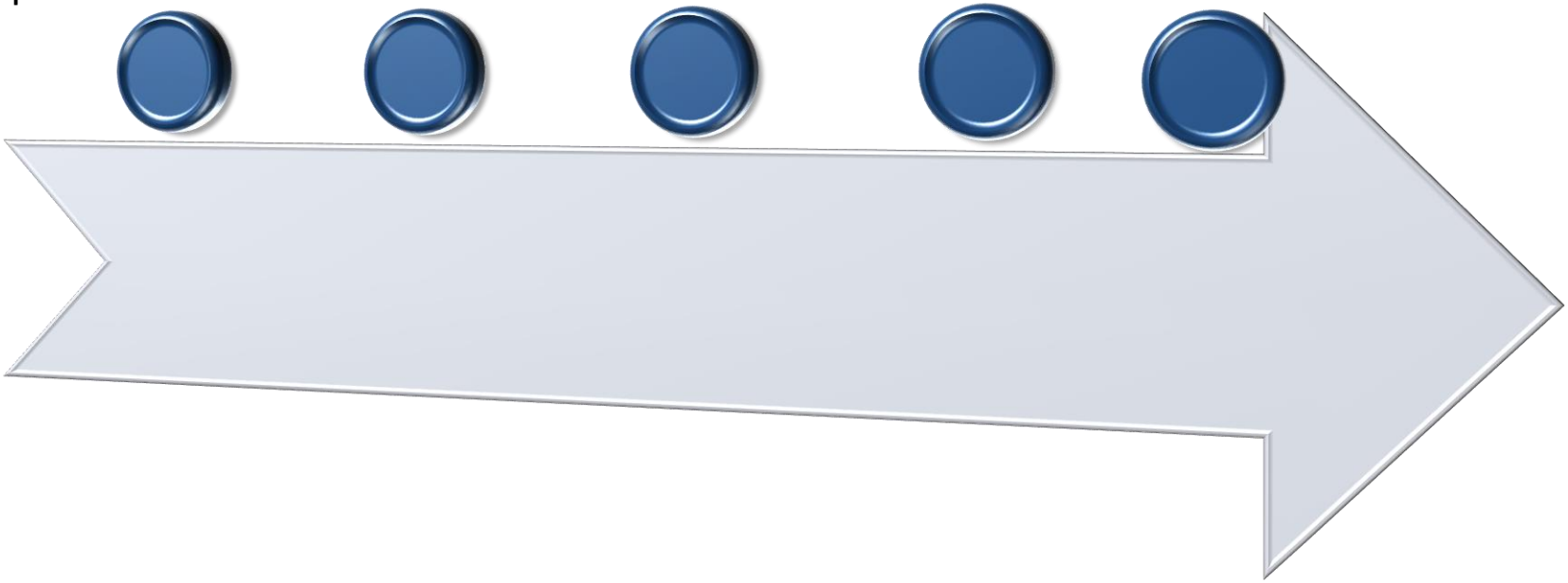
Further potential clinical manifestations include acute renal dysfunction, acid-base disturbances on blood gases, hyperglycaemia, thrombocytopenia and impaired consciousness


Rapid progression to septic shock may occur and may even be the presenting feature in patients with severe emphysematous pyelonephritis.
Loin tenderness is the most common physical sign.
In some cases crepitus around the flank area may also be felt

Laboratory data in 70–80% of the reported cases showed leucocytosis while thrombocytopenia was seen in 15–20% of the patients.

As most of these patients have diabetes, high blood glucose level is a common finding.

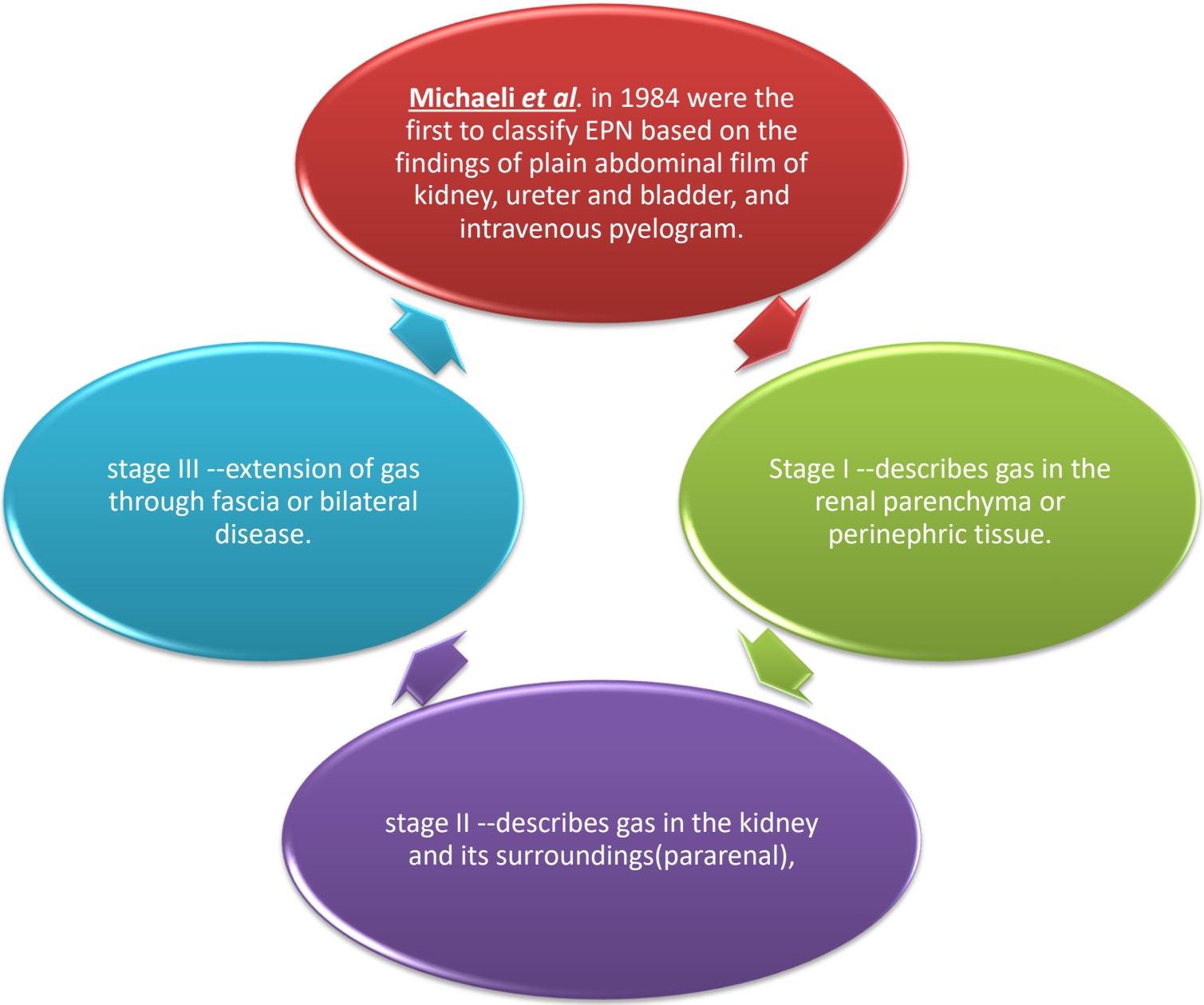
Acute renal failure, microscopic or macroscopic haematuria and severe proteinuria are other positive findings in EPN.



CLASSIFICATION

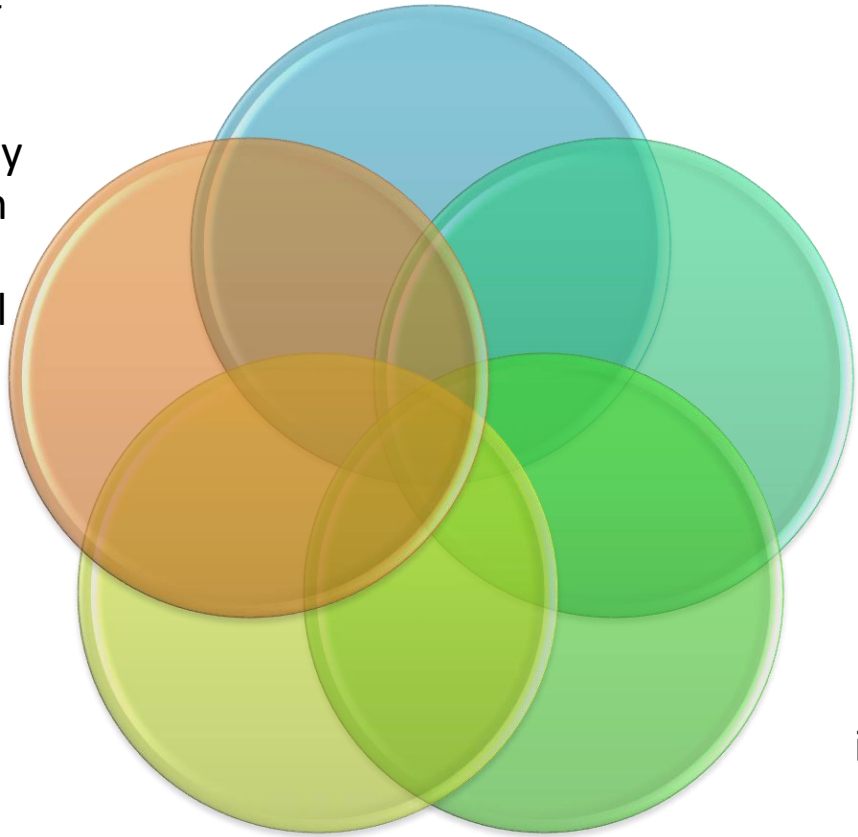
Various classifications exist for EPN.

There are three classifications of EPN based on radiological findings.

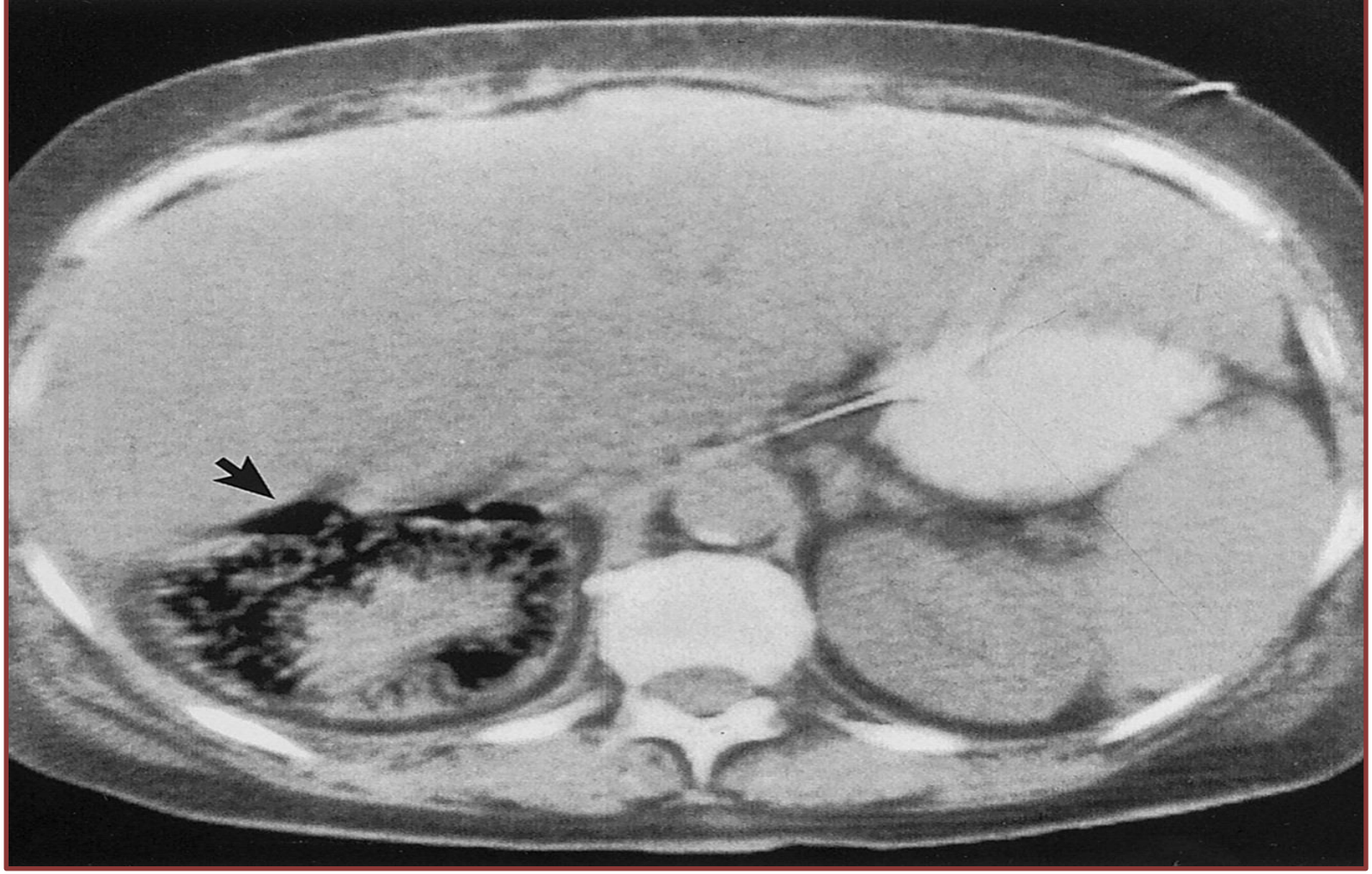

Michaeli *et al*. in 1984 were the first to classify EPN based on the findings of plain abdominal film of kidney, ureter and bladder, and intravenous pyelogram.
Stage I --describes gas in the renal parenchyma or perinephric tissue.
stage II --describes gas in the kidney and its surroundings(pararenal),
stage III --extension of gas through fascia or bilateral disease.

Wan *et al.* in 1996 categorized EPN into two groups based on CT findings.

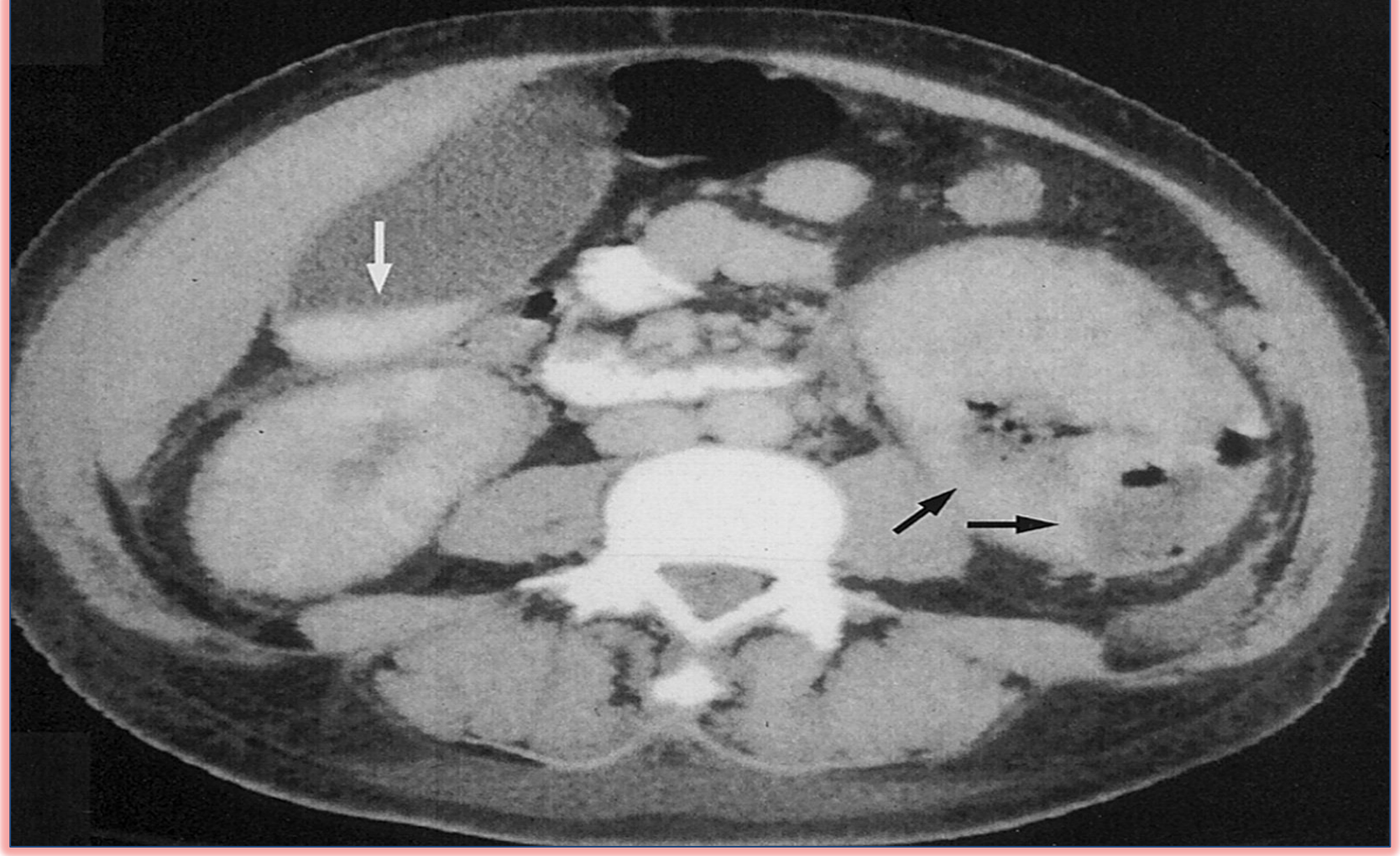
Type II EPN-- either the presence of renal or perirenal fluid in association with a bubbly or loculated gas pattern or gas in the collecting system with acute renal or perirenal fluid-containing abscesses.



Type I EPN-- parenchymal destruction with total absence of fluid content on the CT images and presence of a streaky or mottled gas pattern on CT scan



Type I emphysematous pyelonephritis. Contrast-enhanced CT scan demonstrates gas that radiates diffusely throughout the renal parenchyma (arrow). No associated fluid collections are seen.



Type II emphysematous pyelonephritis. CT scan obtained in a different patient shows several small foci of gas, some with associated regions of fluid attenuation (black arrows). These fluid-attenuation regions are thought to represent a more favorable immune response

Type I emphysematous pyelonephritis has a 65-70% mortality rate versus 15-20% for type II, although transformation from type I to type II has been observed following conservative treatment

In 2000, **Huang and Tseng** published a different classification, which was also based on the CT findings but described in more detail and with more sub-categories than the previous one.

Class 1: gas in the collecting system only;

Class 2: gas in the renal parenchyma without extension to perinephric space;

Class 3A: extension of gas or abscess to perinephric space;

Class 3B: extension of gas or abscess to pararenal space;

Class 4: bilateral EPN or solitary kidney with EPN.

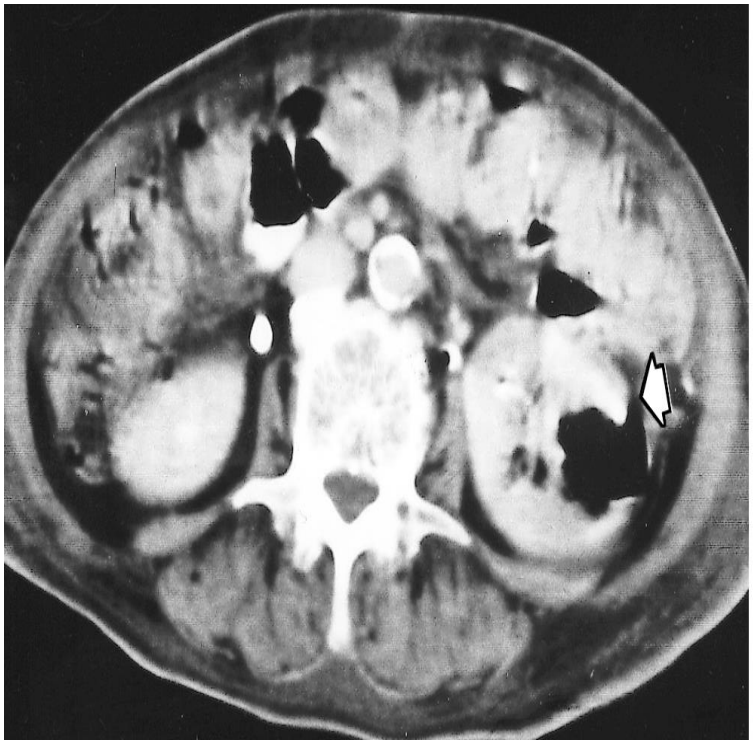
Class 1

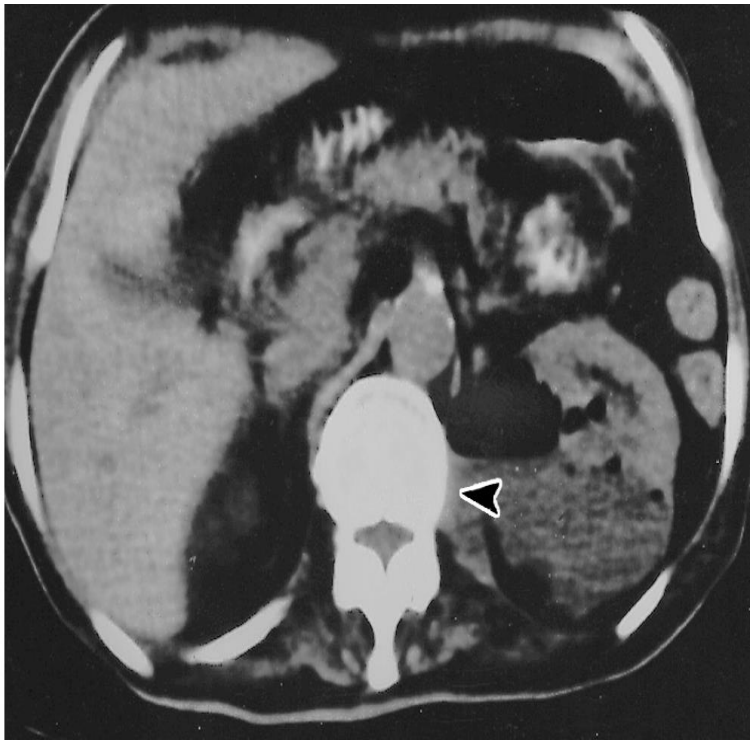
Computed tomographic scan demonstrates gas accumulated only in the right renal pelvis.

A pelvic stone (arrowhead) with hydronephrosis was also found in the right kidney.

Class II

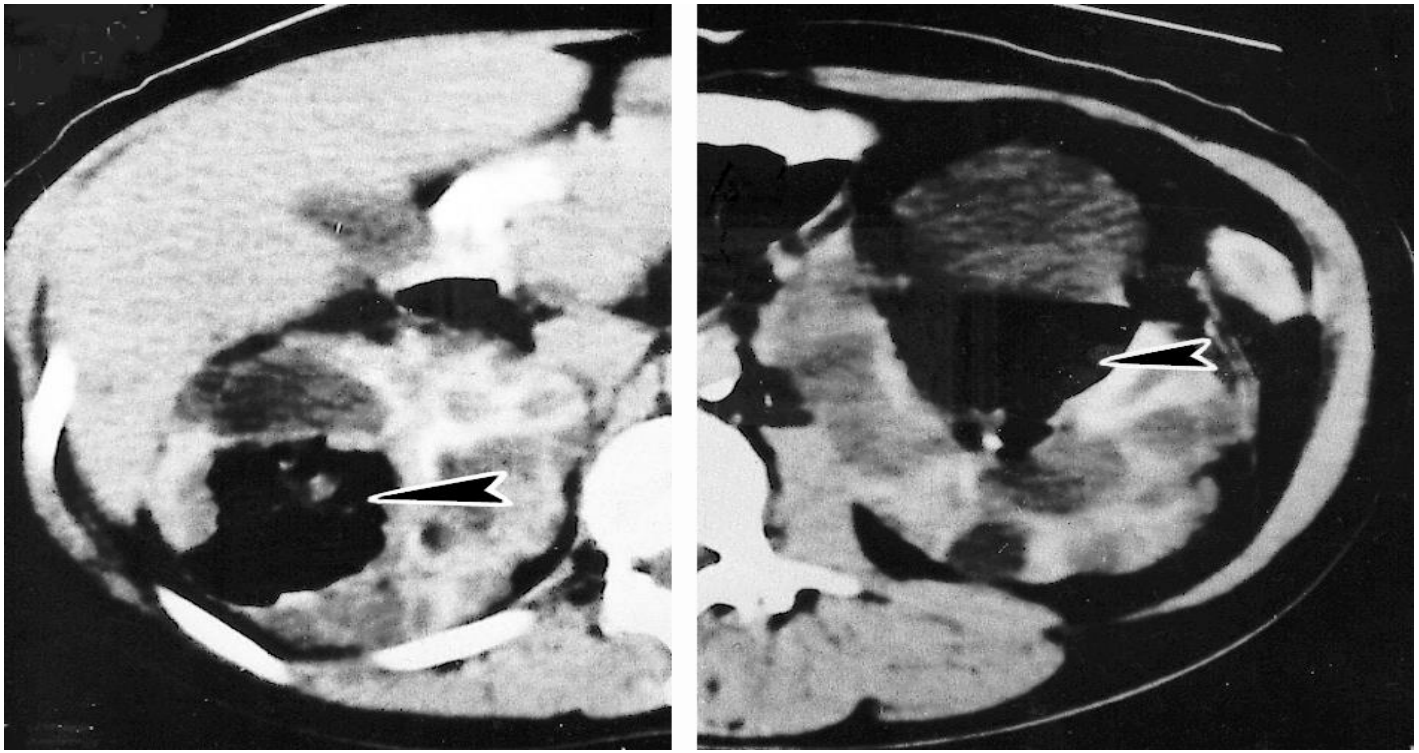
Computed tomographic scan shows gas within the right renal parenchyma and a right renal stone





This computed tomographic scan shows left-sided EPN with extension of gas to the perinephric space(3A) (white arrow).

Right, Class 3B EPN. Computed tomographic scan shows left-sided EPN with extension of gas to the pararenal space (arrowhead)



Class 4

Computed tomographic scan shows accumulation of gas (arrowheads) in both kidneys in a patient with autosomal dominant polycystic kidney disease.

DIAGNOSIS

Emphysematous pyelonephritis is a radiological diagnosis which requires imaging, since most of the clinical and the laboratory findings will only indicate sepsis of renal origin.

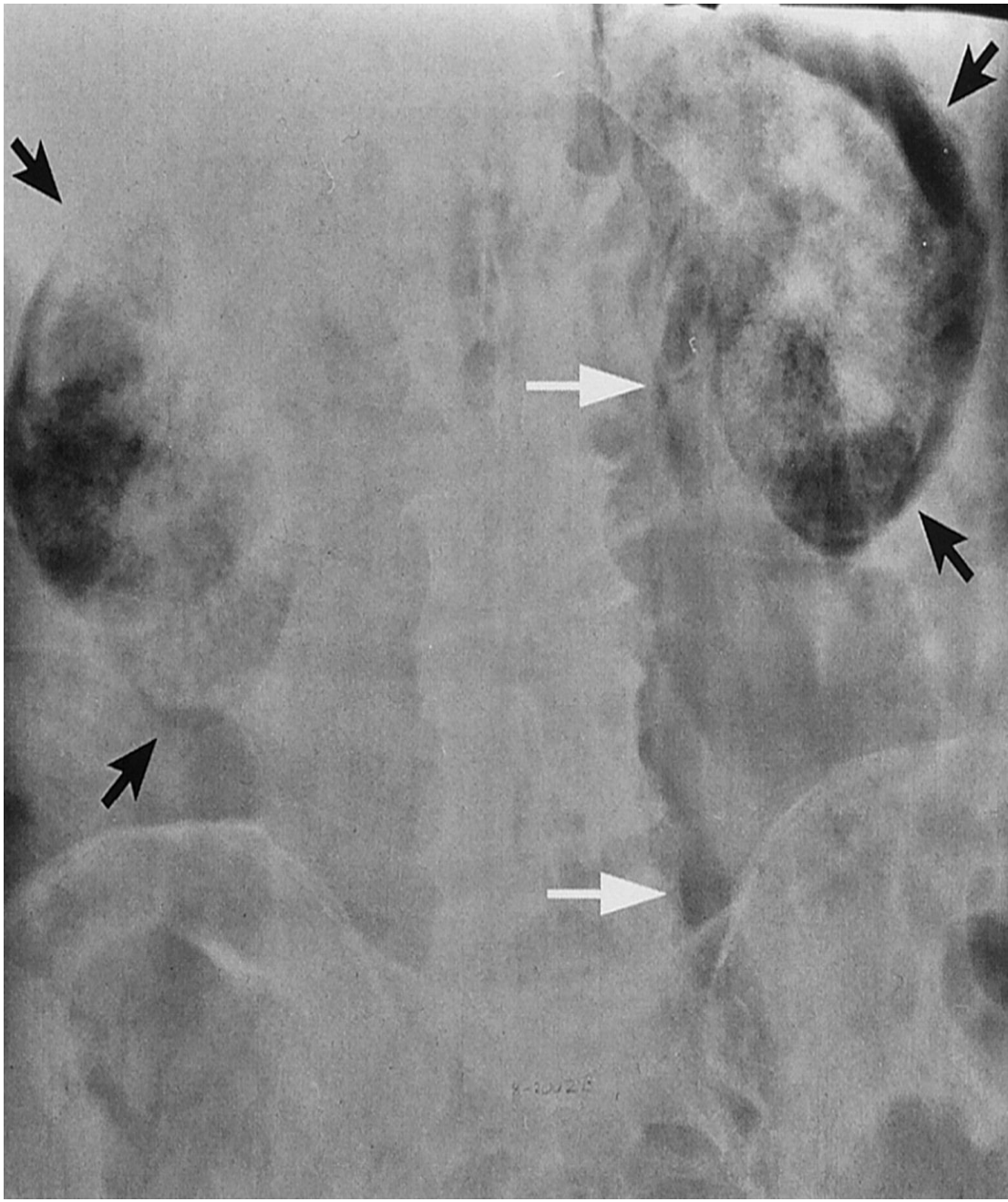
Tissue gas that is distributed in the parenchyma may appear on **abdominal radiographs** as mottled gas shadows over the involved kidney.

This finding is often mistaken for bowel gas.

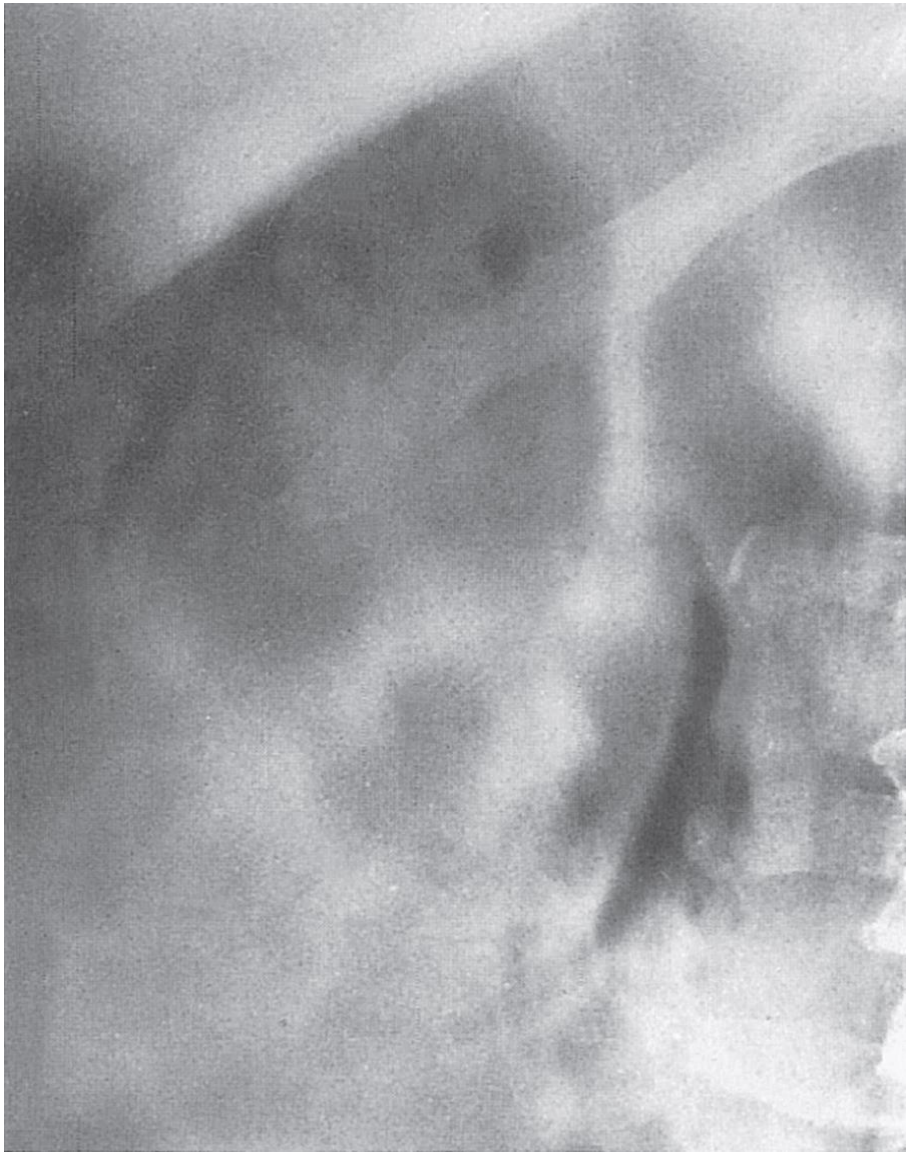
A crescentic collection of gas over the upper pole of the kidney is more distinctive.

As the infection progresses, gas extends to the perinephric space and retroperitoneum.

Abdominal radiograph reveals extensive, radially oriented air within and surrounding the kidneys (black arrows). Air is also seen within the left renal collecting system (white arrows)



Extensive perinephric *and intraparenchymal gas in emphysematous* pyelonephritis

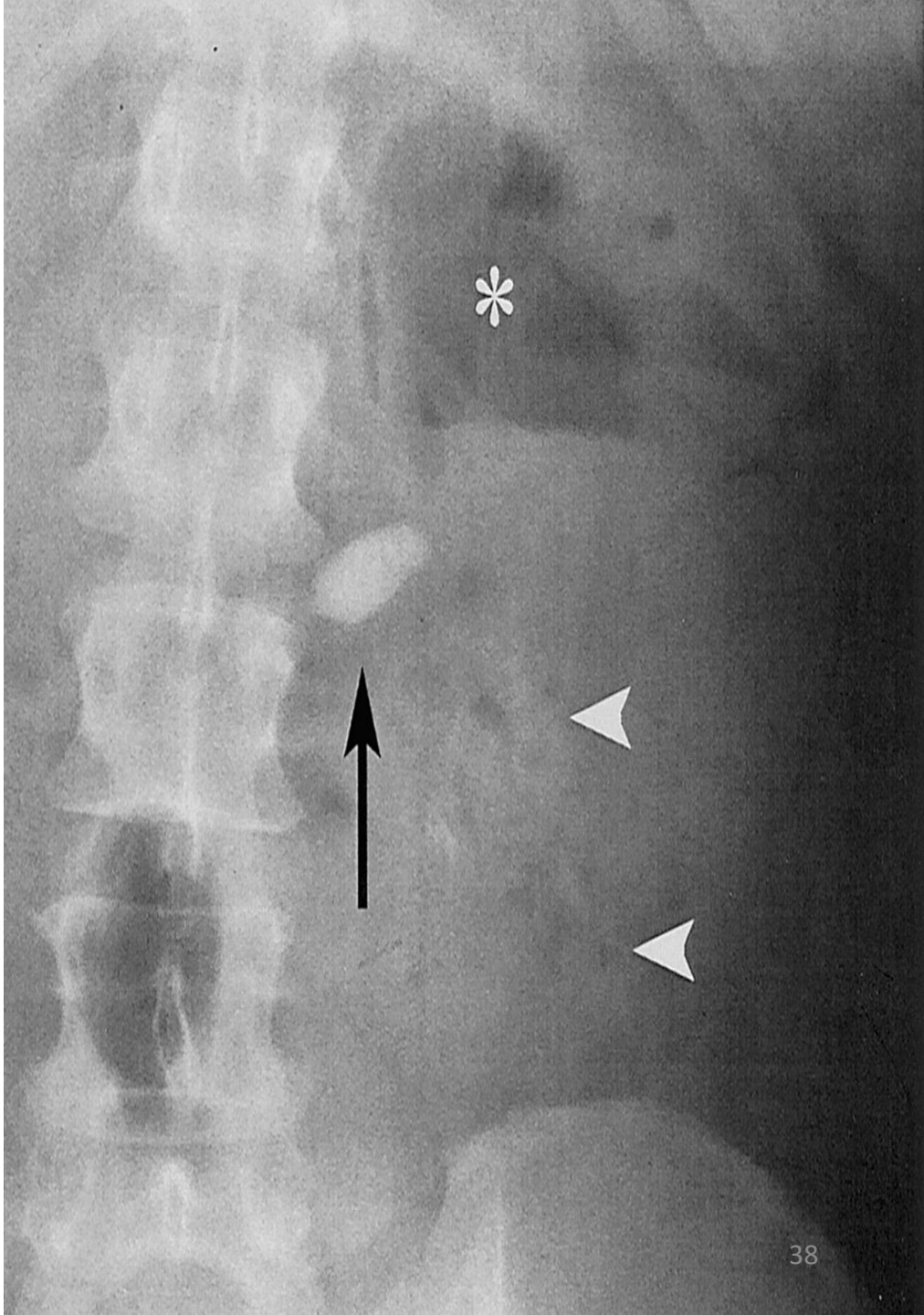


Intravenous urography will demonstrate a persistent nephrogram on the affected side secondary to delayed excretion of contrast material.

The ipsilateral psoas shadow may be obscured and obstructing stones identified at abdominal radiography.

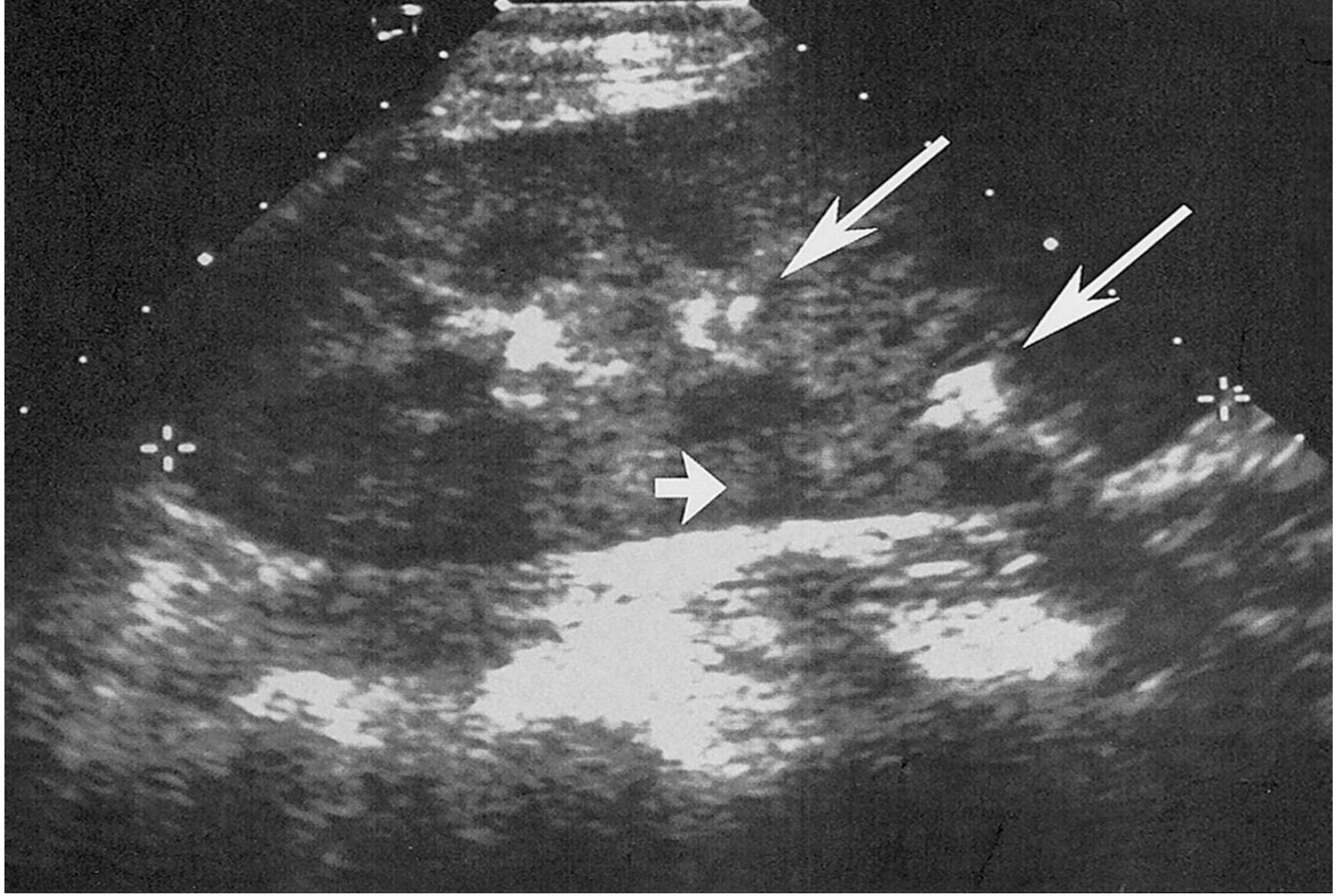
Abdominal radiograph (IVP) obtained with the patient upright demonstrates a 2-cm calcification overlying the region of the left ureteropelvic junction (arrow).

Note also the mottled collection of gas bubbles in the region of the left lower renal pole (arrowheads) and the large air-fluid level within the upper pole (*)



Ultrasonography

US will characteristically show an enlarged kidney containing high-amplitude echoes within the renal parenchyma, however, the depth of parenchymal involvement may be underestimated at US, and multiple renal stones may also manifest as echogenic foci with “clean” posterior acoustic shadowing



US image of the left kidney demonstrates multiple hyperechoic foci (long arrows) with dirty acoustic shadowing (short arrow), a finding that represents gas within the renal parenchyma.

CECT ABDOMEN

Although plain radiography and ultrasound may suggest EPN, computed tomography (CT) of the abdomen is more sensitive, allows for more accurate staging of the disease and is **considered the gold standard for diagnosis.**

Ultrasonography and plain radiograph of the abdomen are only accurate in 59% and 52% of cases, respectively, so abdominal CT is necessary for early diagnosis and further management of EPN.

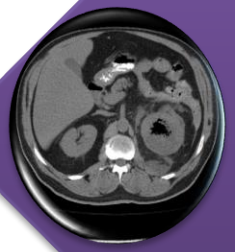
The use of intravenous contrast material will often reveal asymmetric renal enhancement or delayed excretion, and, during the nephrographic phase, will help identify areas of focal tissue necrosis or abscess formation.

A **nuclear renal scan** should be performed to assess the degree of renal function impairment of the involved kidney and the status of the contralateral kidney before going for nephrectomy.

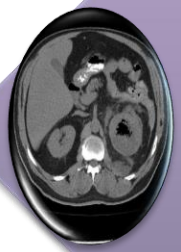
MANAGEMENT

In patients who are being treated for pyelonephritis, the radiological diagnosis may be missed, unless appropriate imaging is obtained.

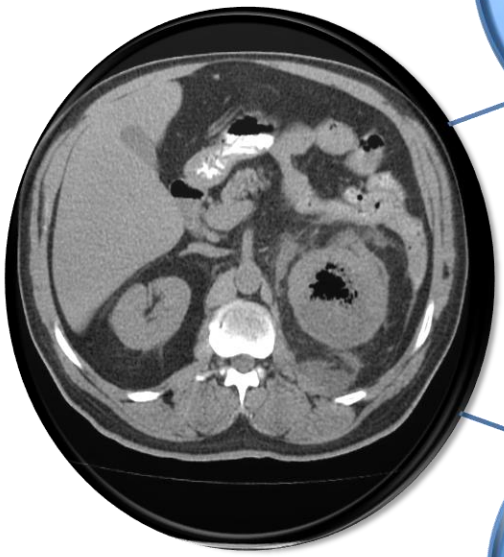
This group of patients, along with those who fail to respond to the standard line of treatment of pyelonephritis, should have an urgent CT scan to confirm the diagnosis.



Basic resuscitation measures of intravenous fluids, acid base balance correction and appropriate antibiotics should be commenced along with good glycaemic control.

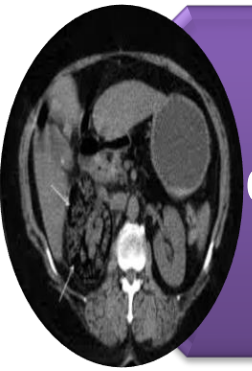


It is important to maintain a systolic blood pressure of more than 100 mmHg, with fluid resuscitation or ionotropic support if required.

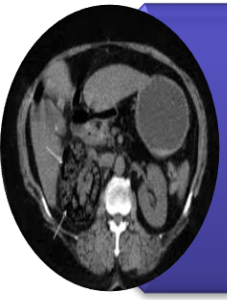


Meta-analysis of the risk factors affecting the mortality rate concluded that a systolic blood pressure of 90 mmHg adversely affected the mortality rate when compared with a pressure of more than 100 mmHg in patients of EPN.

If the clinical condition and the laboratory results show deterioration, then the level of care should be stepped up as these patients may require multi-organ support.

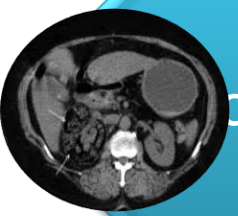


Gram-negative bacteria remain the most common causative organisms so the initial antibiotic regimen should target them.



cephalosporins and quinolones can be used.

A combination of aminoglycoside with any of the other two groups can be used in the initial treatment stage.

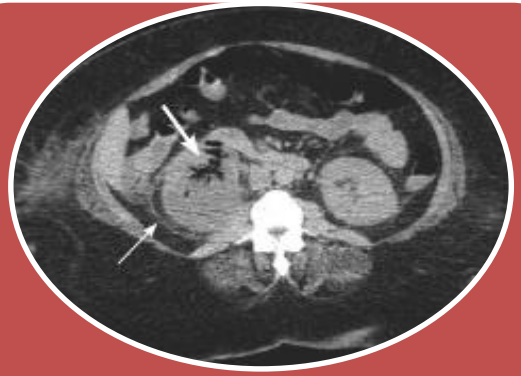


Once the culture report is available, the antibiotics can be changed according to the type of organisms along with their individual sensitivities.

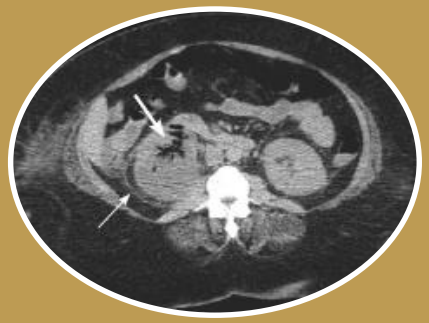
The accepted treatment of EPN until the late 1980s was emergency nephrectomy and/or open surgical drainage together with antibiotic therapy, resulting in a reported mortality rate of 40–50%.

Some of the authors suggested early nephrectomy along with medical management to reduce the mortality rate and shorten the recovery period.

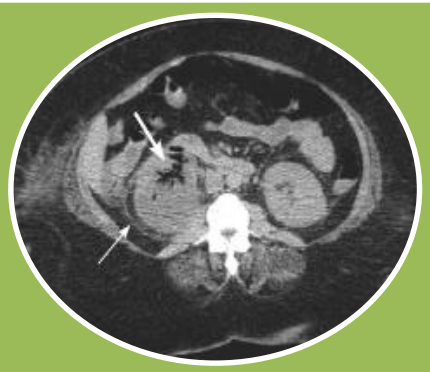
Percutaneous Drainage



Percutaneous drainage(PCD) as a treatment option for EPN, which was first shown by Hudson *et al.*



Subsequent case studies have shown patients being successfully treated with PCD when used in addition to medical management, with significant reduction in the mortality rates.



PCD helps to preserve the function of the affected kidney in about 70-80% of cases.

PCD should be performed on patients who have localized areas of gas and in whom functioning renal tissue is believed to be present.

A pigtail drain of at least 14 Fr in size should be inserted, either with USG or CT guidance but ideally with CT guidance which has a better success rate when compared with an ultrasonography.

An abscess with loculations or multiple abscesses is not a contraindication for PCD, as more than one catheter can be used to drain all loculations.

The abscess, which is technically easier to access and would significantly reduce the pressure on the viable renal tissue, should be targeted first with PCD.

During the last decade there has been a gradual shift toward a nephron-sparing approach with PCD, with or without elective nephrectomy at a later stage.

The treatment strategies include MM alone, PCD plus MM, MM plus emergency nephrectomy, and PCD plus MM plus emergency nephrectomy.

Patients on PCD plus MM benefit from follow-up CT in 4 to 7 weeks as recommended by **Chen *et al.*** to look for air/fluid collections.

This will also be helpful in planning a nephrectomy for non-responders to PCD plus MM.

In a meta analysis of the management strategies, the most successful management was MM with PCD (80–100%), which was also associated with the lowest mortality at 13.5%.

In the small proportion of patients managed with MM and PCD, subsequent nephrectomy will be required and in these patients the reported mortality is 6.6%.

In Class 1 and 2 EPN (based on the Huang and Tseng classification) MM alone or combined with PCD can provide a good outcome.

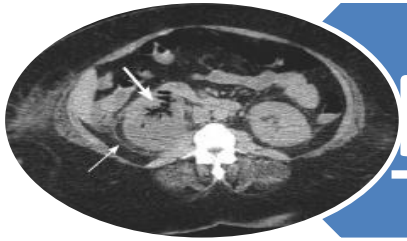
In Class 3 and 4 EPN, MM plus PCD provides a survival rate of 85-92% .

Nephrectomy in patients with EPN can be simple, radical or laparoscopic. Simple nephrectomy can be carried out with a mortality rate as low as 5%.

Laparoscopic nephrectomy can be successfully performed in these patients and has the advantage of providing a shortened recovery period and hospital stay.

In addition patients with EPN with coexisting nephropathy might need renal support measures in the form of dialysis and a recent study concluded that the availability of renal support seems to reduce the mortality rate.

The long-term outcome for renal function and the need for further support will depend on the degree and amount of parenchymal loss and coexisting renal disease.



PROGNOSTIC FACTORS

Emphysematous pyelonephritis requires urgent attention because of the life threatening potential associated with the septic complications.

Various risk factors have been identified and reported in the literature.

The presence of **diabetes mellitus** appeared to be a common risk factor for EPN but, surprisingly, it is not associated with increased mortality.

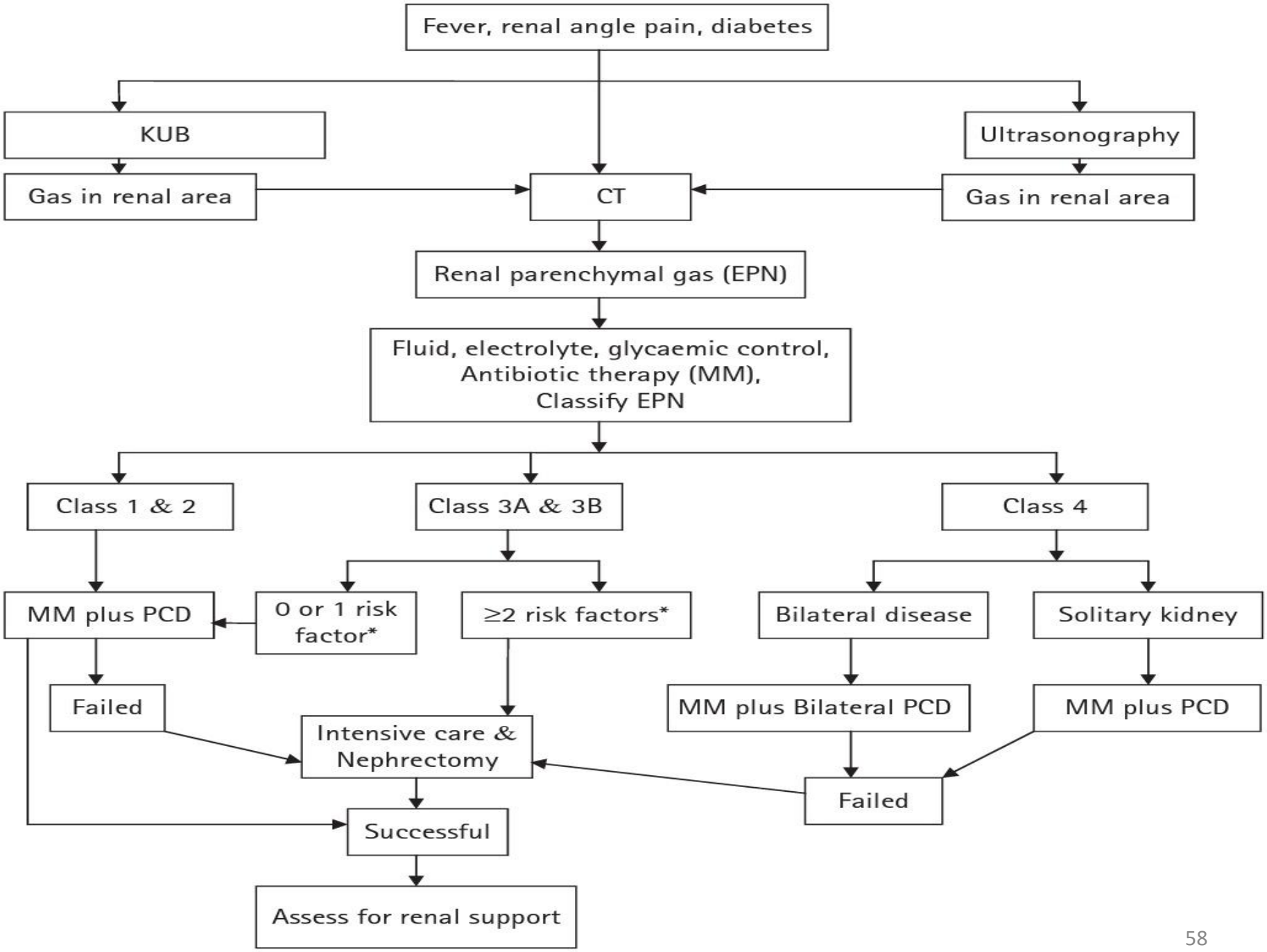
Significant association could be established between higher mortality in EPN with

- nephrolithiasis,
- *E. coli* or *K. pneumoniae* aetiology of EPN,
- age >50 years,
- female sex,
- history of UTIs,
- alcoholism.

Systolic blood pressure less than 90 mmHg, disturbance of consciousness as well as increase in serum creatinine level were found to be associated with higher mortality also.

The presence of thrombocytopenia and bilateral EPN are both linked with poor prognosis.

MM with antibiotics alone is associated with a higher risk of mortality when compared with additional interventions of percutaneous drainage (PCD) of the abscess or nephrectomy


Fever, renal angle pain, diabetes
KUB
Ultrasonography
Gas in renal area
CT
Gas in renal area
Renal parenchymal gas (EPN)
Fluid, electrolyte, glycaemic control,
Antibiotic therapy (MM),
Classify EPN
Class 1 & 2
Class 3A & 3B
Class 4
MM plus PCD
0 or 1 risk factor*
≥2 risk factors*
Bilateral disease
Solitary kidney
Failed
MM plus Bilateral PCD
MM plus PCD
Intensive care & Nephrectomy
Failed
Successful
Assess for renal support

In conclusion, EPN is a potentially life threatening condition which is most commonly associated with poorly controlled diabetes.

It requires a high index of suspicion in patients not responding to the routine management of pyelonephritis.

It is a radiological diagnosis and CT is the best investigation.

Aggressive resuscitation should be done and the condition is currently treated by MM along with PCD mostly commonly.

Some patients may not respond and emergency nephrectomy may be required.

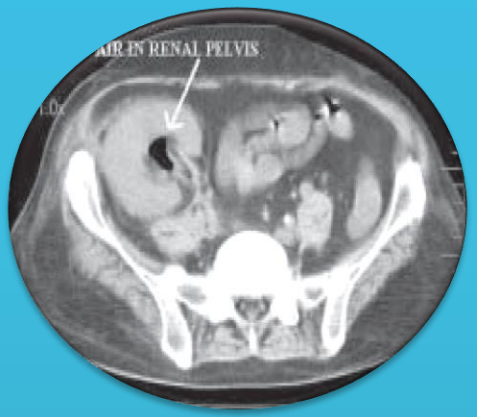
Reported mortality figures have improved since the 1970s but still are at 20-25%

Emphysematous Pyelitis

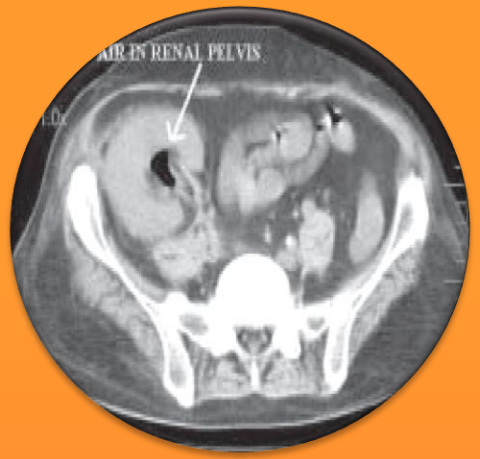
Emphysematous pyelitis is the term used to describe the presence of gas limited to the renal excretory system.

Although the urothelium may be primarily involved, the gas is usually secondary to coexistent bacterial infections of the kidney or urinary bladder, with *E coli* being the most commonly cultured bacteria.

Emphysematous pyelitis is seen more commonly in women, is often associated with underlying diabetes or obstructing stone disease, and carries a mortality rate of up to 10%, which is significantly lower than that of emphysematous pyelonephritis.



At conventional radiography, gas is seen filling and outlining the ureters and pelvicaliceal system.



CT best delineates gas within the collecting system and helps reliably identify ureteric stones. More importantly, CT helps exclude complicated forms of emphysematous pyelitis, such as the presence of renal or perirenal fluid collections, frank abscesses, or emphysematous pyelonephritis.

Potential noninfectious sources of gas within the collecting system that should be excluded which includes reflux of air during instrumentation and the presence of an ureterosigmoidostomy.

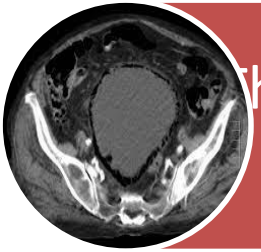
In the absence of obstruction, surgical or percutaneous drainage procedures are rarely needed, and emphysematous pyelitis will likely respond to intravenous antimicrobial therapy and careful attention to underlying co-morbid conditions.

The persistence of gas following conservative therapy indicates continued infection, which may then require more aggressive intervention.

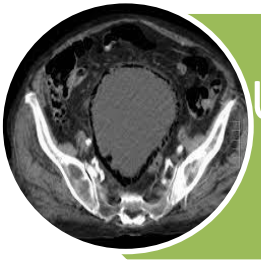
Emphysematous Cystitis

Emphysematous cystitis represents a rare form of acute inflammation of the bladder mucosa and underlying musculature.

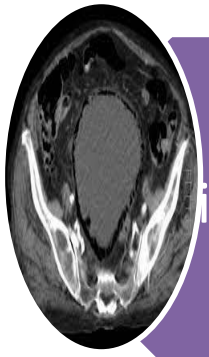
Clinical symptoms of dysuria, increased urinary frequency, and hematuria are common.



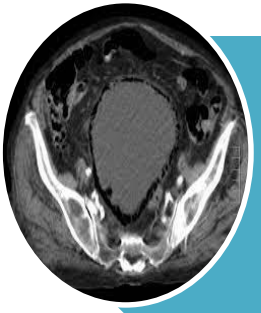
The presence of pneumaturia is a rare, although more specific, clinical finding.



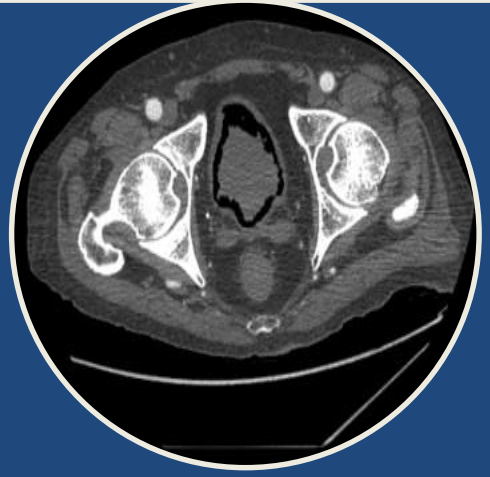
Underlying diabetes mellitus is present in over half of reported cases, with women being affected twice as often as men.



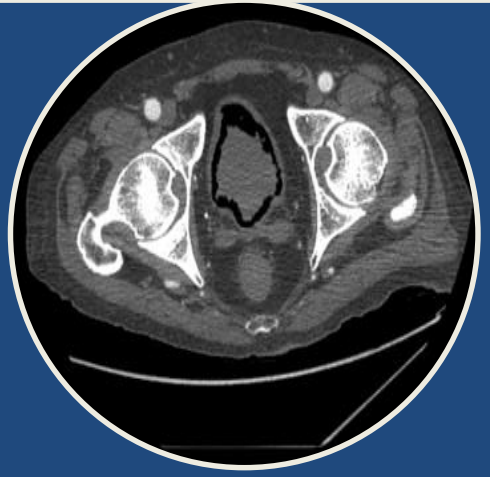
Other predisposing conditions include chronic urinary tract infections, bladder outlet obstruction, and a neurogenic bladder (an increasingly common complication in elderly diabetic patients).



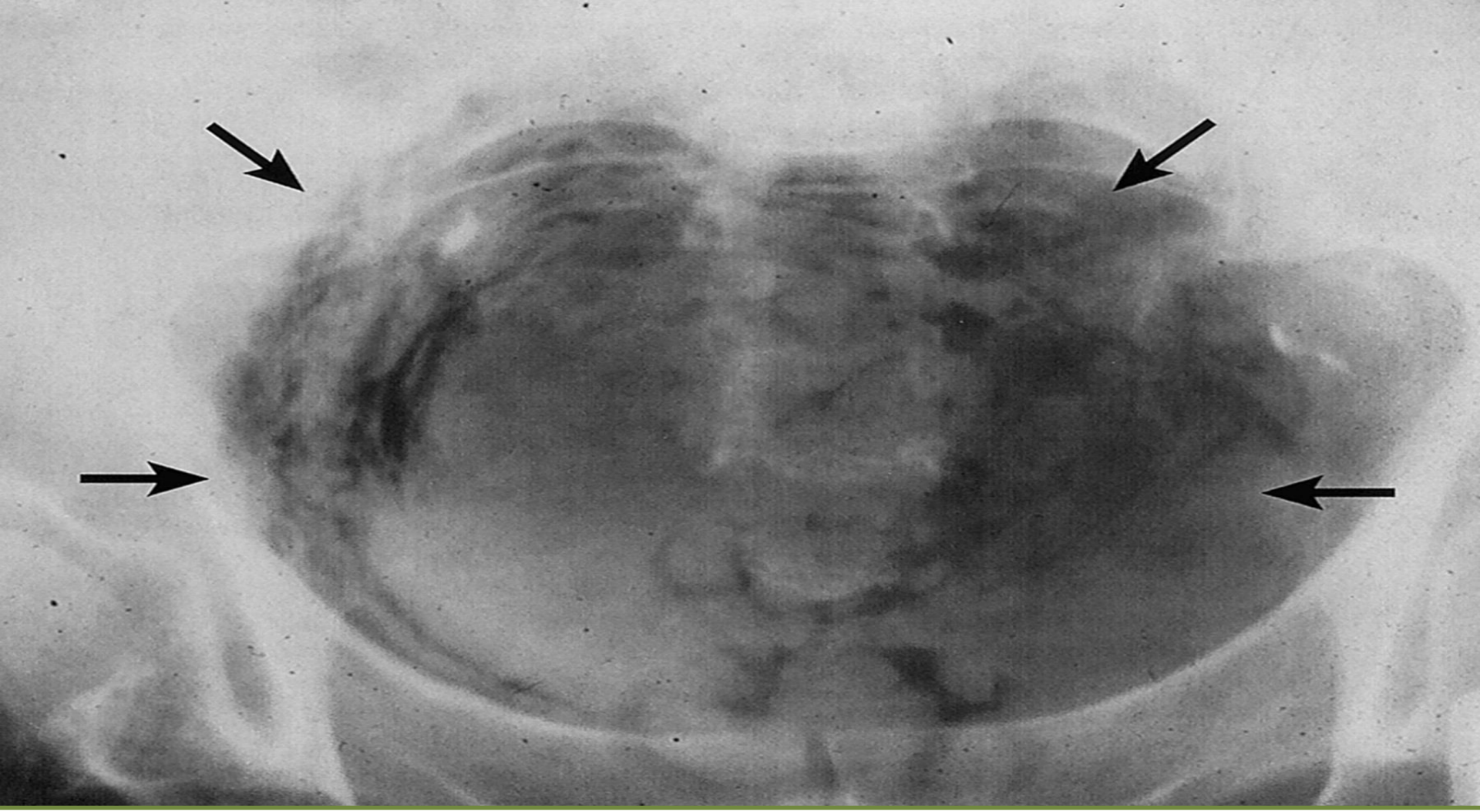
Frequently, isolated gas-producing bacteria include the coliform bacteria *E coli* and *Enterobacter* aerogens, although *Clostridia* and fungal species are occasionally identified.



Possible noninfectious sources of pelvic air should be considered and include recent bladder instrumentation, vesicocolic or vesicovaginal fistulas, trauma, and pneumatosis cystoides intestinalis.



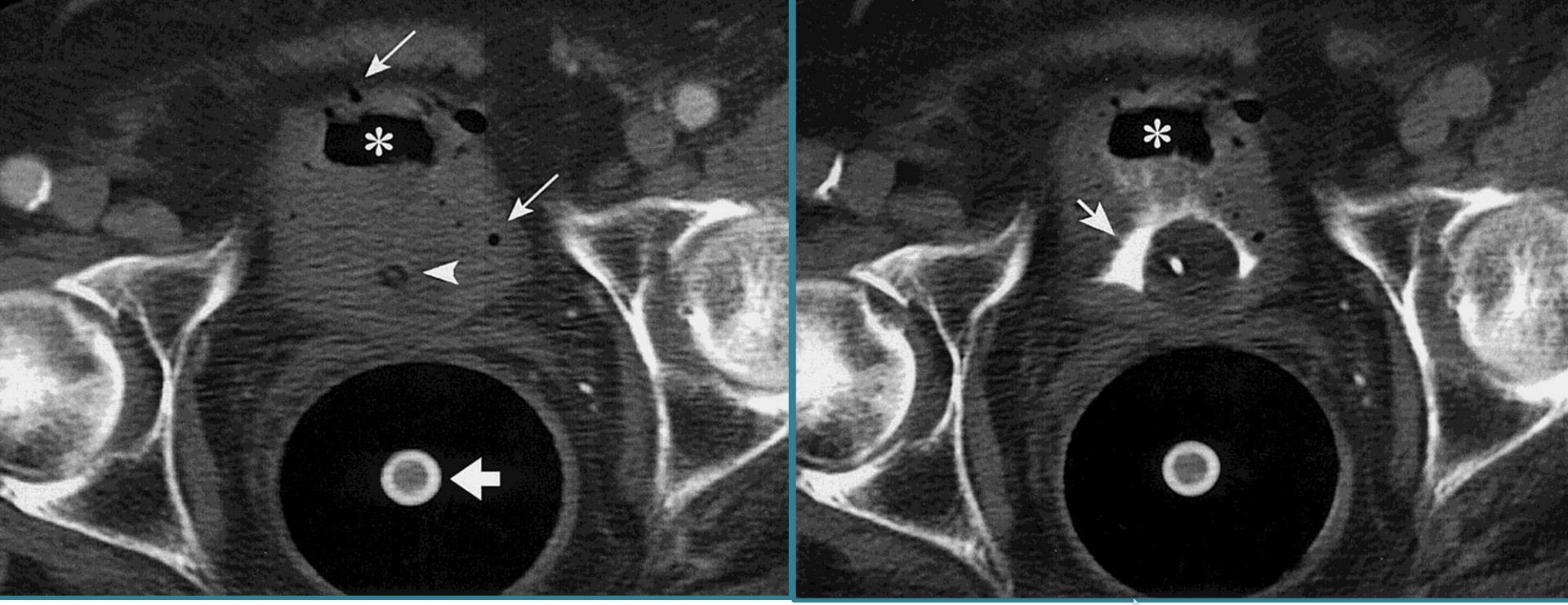
Conventional radiography of emphysematous cystitis characteristically shows curvilinear or mottled areas of increased radiolucency in the region of the urinary bladder, separate from more posterior rectal gas.



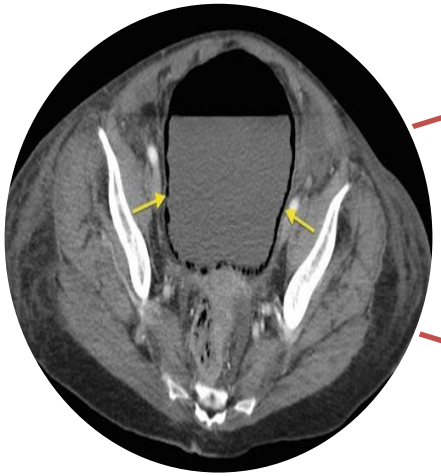
Pelvic radiograph obtained with the patient upright demonstrates circumlinear streaks of increased radiolucency representing air in the expected location of the urinary bladder (arrows).

US will commonly demonstrate diffuse bladder wall thickening and increased echogenicity. Focal regions of high-amplitude echoes with posterior dirty acoustic shadowing into the lumen may be seen in extensive cases.

CT is a highly sensitive examination that allows early detection of intraluminal or intramural gas.

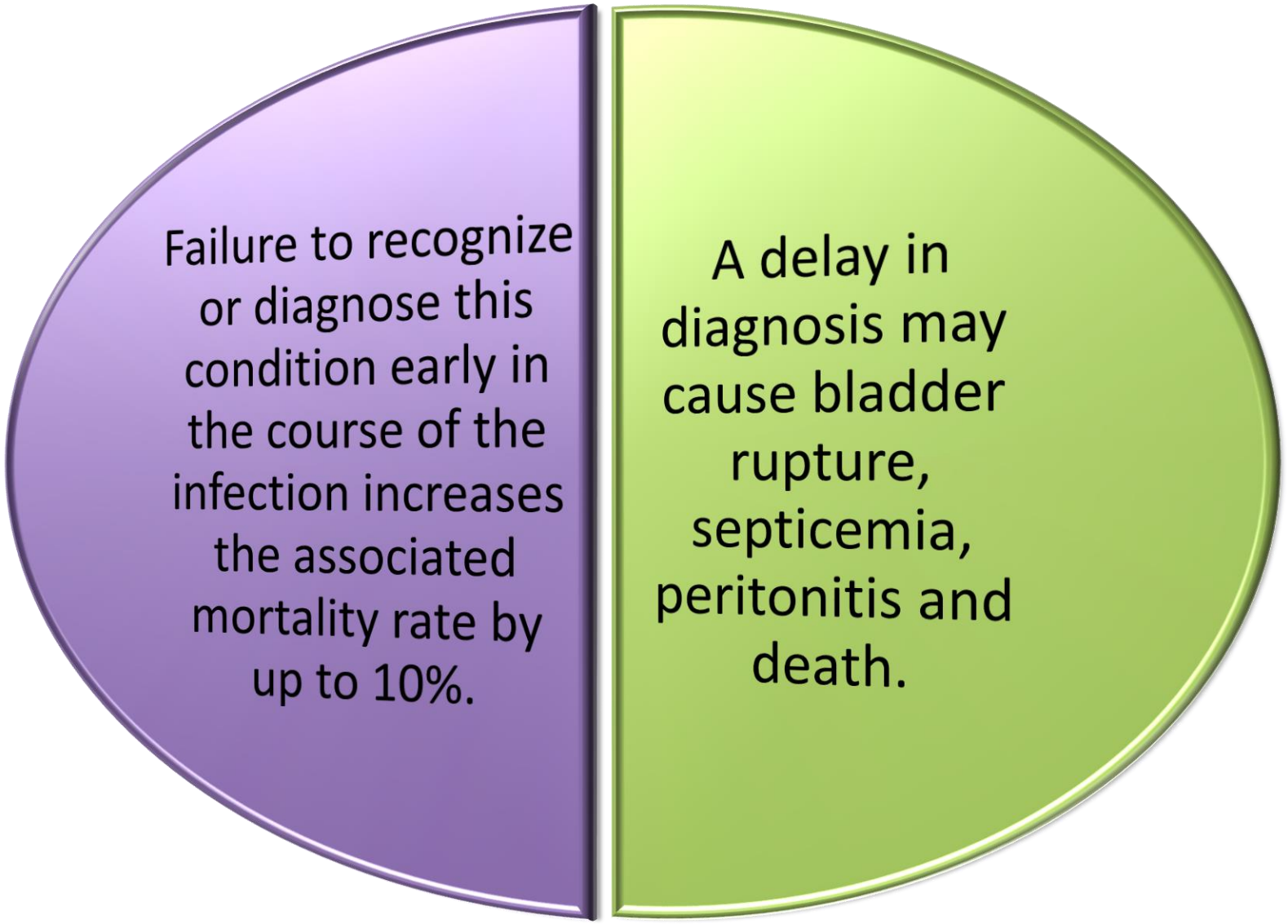


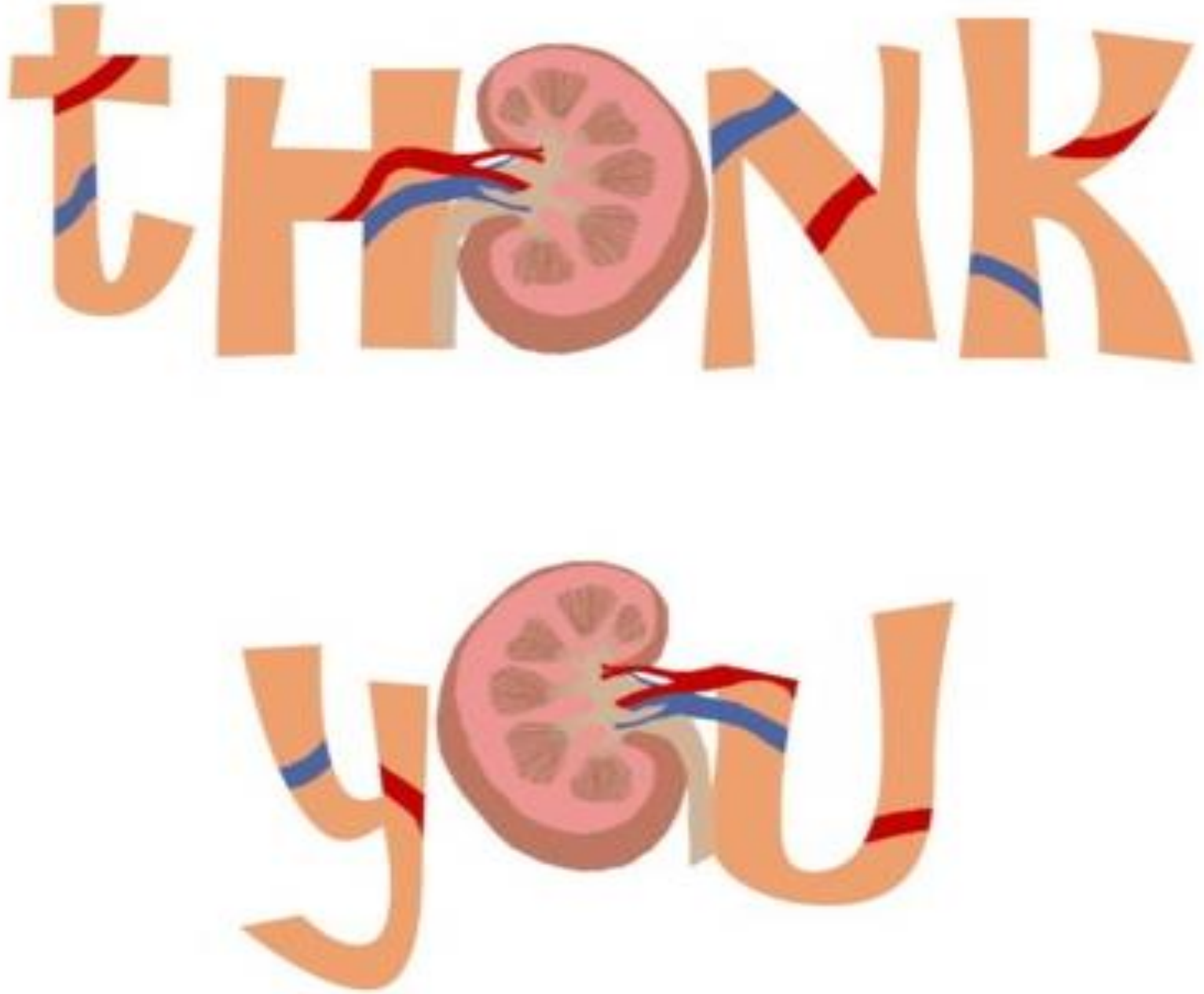
Emphysematous cystitis in an 81-year-old diabetic man undergoing treatment for acute pancreatitis. **(a)** Contrast-enhanced arterial-phase CT scan through the pelvis shows a Foley catheter (arrowhead) and a rectal tube with a balloon cuff (thick arrow). There is a rounded collection of air within the urinary bladder (*) as well as multiple smaller locules of air more peripherally (thin arrows). **(b)** Delayed CT scan obtained at the same level shows contrast material within the bladder lumen (arrow). Both the large (*) and small collections of gas are clearly seen to be within the thickened anterior bladder wall rather than the bladder lumen



Treatment for emphysematous cystitis involves broad-spectrum antimicrobial therapy, hyperglycemic control, and adequate urine drainage with correction of possible bladder outlet obstruction when present.

Surgical interventions, such as debridement or partial cystectomy, may be required for patients who respond poorly to antibiotics or have necrotizing tissue.


Failure to recognize or diagnose this condition early in the course of the infection increases the associated mortality rate by up to 10%.
A delay in diagnosis may cause bladder rupture, septicemia, peritonitis and death.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