

Chyluria

- Chyluria is a state of chronic lymphourinary reflux via fistulous communications secondary to lymphatic stasis caused by obstruction of the lymphatic flow.
- **Lymphatic drainage** of the kidney occurs in a trilaminar fashion.

The 1st lamina= lies within the renal parenchyma,

the 2nd= lies at a sub-capsular level and

the 3rd= lies within the perinephric fat.

The intrarenal lymphatics emerge as 4-7 trunks, which emerge at the renal hilum to join the 2nd and 3rd level lymphatics.

These then eventually converge along the renal vessels to the lateral aortic nodes.

Physiologically =

Chyle travels from the lacteals to the cisterna chyli or thoracic duct.

Pathological obstruction and/or insufficiency of the valvular system of lymph

channels leads to retrograde flow to lumbar lymph glands draining renal lymphatics. Thus there is a short

circuiting of chyle drainage from intestinal
lacteals to renal lymphatics

etiology :

Chyluria occurs after rupture of lymphatic varices into renal tubules.

The lymphatic varices are the result of high intralymphatic pressure, usually due to an obstruction or stenosis of the major lymphatic ducts.

Chyluria is associated with abnormal retrograde or lateral flow of lymph from the intestinal lymphatics to the kidney, ureter or bladder allowing chylous material to be discharged into the urinary collecting system

Various causative factors have been implicated.

Passage of chyle into the urine has been related to a consequence of the rupturing of the lymphatic varices leading to rupture of one or more perirenal lymphatic vessels to the pyelocaliceal system, characterized by milky urine, rich in protein, lipids, cholesterol and triglycerides.

A malformation of the chyliferous vessels of the small intestine associated to hypoplasia of Pecquet's cistern has also been suggested.

- **1)Parasitic (primary-tropical)**

Wuchereria bancrofti (90%)

Taenia echinococcus

Taenia nana

Ankylostomiasis

Trichiniasis

Malarial parasites

- **2)nonparasitic**

Congenital

Lymphangioma of urinary tract

Megalymphatics & urethral/vesical fistulae

Stenosis of thoracic duct

Retroperitoneal lymphangiectasia

Traumatic lymphangiourinary fistulae

Obstruction of thoracic duct/ lymphatics (tumor, granulomas, aortic aneurysm)

Other causes (pregnancy, diabetes, abscess)

Nephrotic syndrome, TB

c/f:

- **The most** common symptom = passage of milky urine in up to 70% cases .
- Other symptoms = **chylous clot, bloody and milky urine, dysuria, frequency, urgency and urinary retention .**

Constitutional symptoms =**weight loss, low-back ache, fever, chills and hydrocele.**

The duration of symptoms may be as long as 2- 11 years .

higher in males (86%) than in females (14%) .

- more frequently reported on the **left side** .
- Majority of the cases present in the 2nd and 3rd decade of life.
About 70% cases have genital manifestations, 25% lymphatic obstruction in limbs and 5% suffer from manifestations like cellulitis, abscesses, hematuria and chyluria.

Grade :

Mild chyluria (34-50%):

Intermittent milky urine; no clot colic/chylous coagulum/urinary retention/weight loss; involvement of single calyx on RGP.

- **Moderate chyluria (33- 40%):**

Intermittent continuous milky urine with occasional clot colic/chylous coagulum; no urinary retention/weight loss; involvement of 2 or more calices on RGP.

- **Severe chyluria (15- 26%):**

Continuous milky urine with clot colic/chylous coagulum/urinary retention /weight loss; involvement of most of calices with/without involvement of ureter on RGP .

INVESTIGATION :

1)Urinary Examination=

A fatty diet a day or night before enhances chyluria and helps in its diagnosis.

≈ A fresh sample of urine is grossly inspected.

≈ The classical urine in chyluria is like=
milk , frequently containing a semisolid gel, blood and fibrin clots.

On standing in test tubes=

it separates into a *top fat layer, middle fibrin and bottom layer of cells and debris.*

Microscopic examination of the sediment comprises of
RBC and lymphocytes

- **Ether test=**

Vigorous shaking (few minutes) with equal amount of ether clears opacity.

- **Methylene blue test=**

Fresh stain with one/two drops of 1:1500 reveals small lymphocytes (single/clump).

- **Sudan III test=**

Oral ingestion of fat labelled with Sudan III (10 gm butter + 100 mg Sudan red III) causes orange pink colouration of urine in chylurics within 2-6 hours

- **2) Biochemistry=**

Urinary triglycerides are invariably present in morning sample, *100% sensitive/specific test*.

- Postprandial urine lipids, especially triglycerides (TGs) ranging from *10 to 1955 mg/dl*, might be used as *markers* for the clinical evaluation of chyluria.
- Urine albumin is abnormally *high* in most cases .

- **3) Immunoelectrophoresis=**

Shows globulins of various types of *apolipoprotein A 48* (of intestinal origin).

- **4) Cystourethroscopy=**

to localize the side of milky efflux from one/both ureteric orifices. *≈ Rarely chylous efflux may be seen from bladder or even posterior urethra.*

≈ A fatty diet previous day/night enhances results.

≈ Ureteric catheterisation (5-6 F) and split urinalysis may be done simultaneously

- **5) RGP=** Demonstrate pyelolymphatic backflow.

not specific as the same is also seen in normal kidneys if contrast is injected under pressure.

≈ 20° Trendelenberg position is used under fluoroscopy for ureteric catheterization to reach renal pelvis

after cystoscopically identifying laterality of efflux.

- Gravity propagated contrast instillation prevents sudden distension of renal pelvis which may lead to loin pain due to opening up of pyelovenous/pyelosinus channels thus causing inadvertent reactions.

- **6)Lymphangiography=**

preoperative imaging procedure of choice and the most powerful diagnostic tool as it demonstrates the site, the caliber and the number of the fistulous communications.

It can also diagnose vascular dysplasias of the lymphatic vessels .

Findings= lymphatico-urinary fistulae at level of kidney, ureter or bladder;

tortuous dilated lymphatics around hilar region (lymphangiectasia) communicating with paravertebral lymphatics; contrast outlining major/minor calyces.

- Other findings = tortuosity and beading of thoracic duct, round granular enlarged

nodes in paraortic area, skipping of lymphatic chain,

dilated cisterna chyli and abnormal lymphatics coursing down along the ureters. In 40% cases contrast may enter PCS and drain via bladder.

not routinely done as it is time consuming, technically demanding and invasive

- Unilateral pedal lymphangiography can detect

lymphaticorenal fistulae via lymphatic crossover even when it is on the side opposite from which contrast agent has been injected.

The advantages of unilateral lymphangiography over bilateral procedures are that it is easy to identify crossover channels, and the discomfort for the patient is reduced because there are fewer incisions and it is quicker

LYMPHANGIOSCINTIGRAPHY:

- it is not as precise as lymphangiography.
- it is a useful, noninvasive, safe, simple technique using Tc99 human serum albumin sulphur colloid, Tc-99m diethylenetriamine pentaacetic acid, 99mTc-antimony sulfide colloid or dextran.

It is **the investigation of choice** (if available) to localize, lateralize and to know the functional extent of reflux and to detect recurrence on follow-up. It can demonstrate abnormal lymphatic drainage in chylurics and allows functional assessment of lymphatic transport and depiction of regional lymph nodes.

A rapid and non-traumatic investigative procedure, it has no known side effects

- **8) Immunological Studies**

ELISA Test: =

useful in diagnosing cases where causative agent is suspected to be filarial and is based on humoral immune response of host to filarial antigen.

≈Filarial IgG antibodies against microfilarial excretory-secretory (mf ES) antigen =

detected in 89% of cases with genital manifestations, 87% with lymphoedema, 67% with lymphadenitis and 60% with other clinical manifestations.

≈ 85% specificity and 95% sensitivity, (it is positive in 3% endemic normals)

- **9) Ultrasonography** : =Echolucent areas seen within blood clot due to the chylous component; leading to heterogenous character of clot.
- **10) Intravenous pyelography** : = Rarely used to demonstrate dilated para-calyceal lymphatics , it may delineate increased renal size in severe disease state due to obstruction.

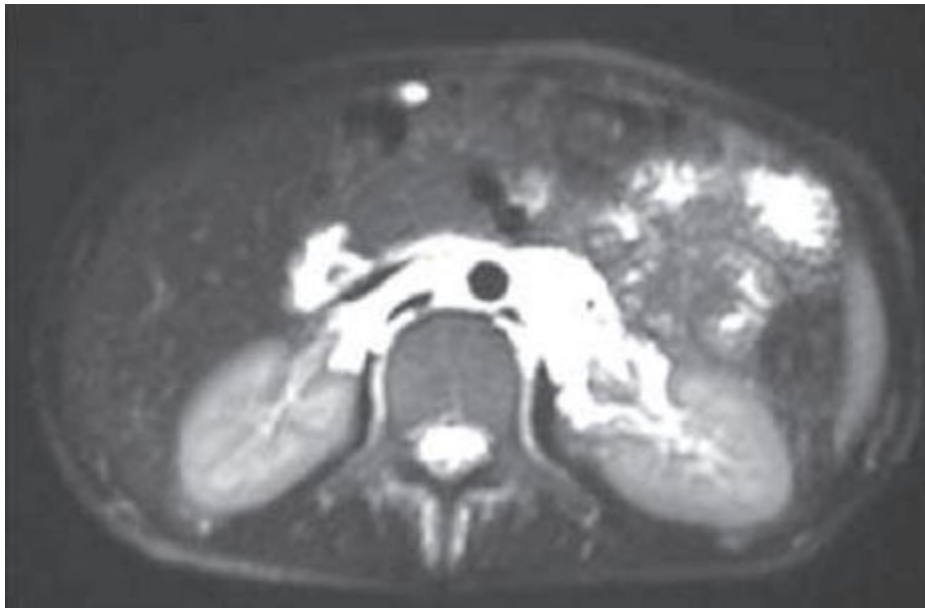
Overall it is not a cost effective investigation

- **11) CT scan** := Fat in the bladder secondary to chyluria picked up on CT .
Dilated lymphatic channels may mimic a mass of confluent low-density lymph nodes
- **12) MR scan** = : -best to delineate anatomy (but no functional assessment).

– most useful in cases where site of obstruction is lower ureter/bladder.

On *MR urography*, the lymphatic channels are seen as *a meshwork of multiple tubular, tortuous, fluid filled structures in the retroperitoneum of the abdomen and pelvis.*

On axial T1W images, these channels are seen as numerous, interconnected small, nodular and streaky intensities and as a cloak of diffuse homogenous hyperintensity.



- **13) Renal biopsy** : undertaken for academic research generally shows immune complex type of glomerulonephritis .

- Others : = Serum type I collagen and type III procollagen are *decreased* in patients with filariasis.
- In contrast, serum hyaluronan (linked to perilymphatic granulomatous inflammation) and serum eosinophil cationic protein is significantly *increased* in patients of filaria exhibiting chyluria

MANAGEMENT:

-) **Non surgical management=**

Dietary modifications: As absorption of dietary fats leads to chyle formation, hence a fat restricted diet is recommended (<25 g/day).

Fats containing MCT (<12 C atoms) are absorbed directly through portals- hence use of coconut oil recommended .

In heavy chyluria= parenteral administration containing fat as MCT with albumin is given.

TPN with enteric rest is advised in intractable chyluria .

- **DEC=** dose is 6mg/kg in 3 divided doses after food for 10-14 days
- **Ivermectin=** 400mg/ kg (single dose) and/or

- **Albendazole**= 400mg may be given along with DEC as symptomatic treatment where filarial infection seems to be the cause.
- *Supportive treatment*:= High protein diet is advised to make up for the albumin lost in the form of chyle.
- *Hematinics, multivitamins and green leafy vegetables* are recommended in hematochyluria.

Bed rest, anti-inflammatory, analgesics and antipyretics = useful in managing in *associated lymphadenitis*.

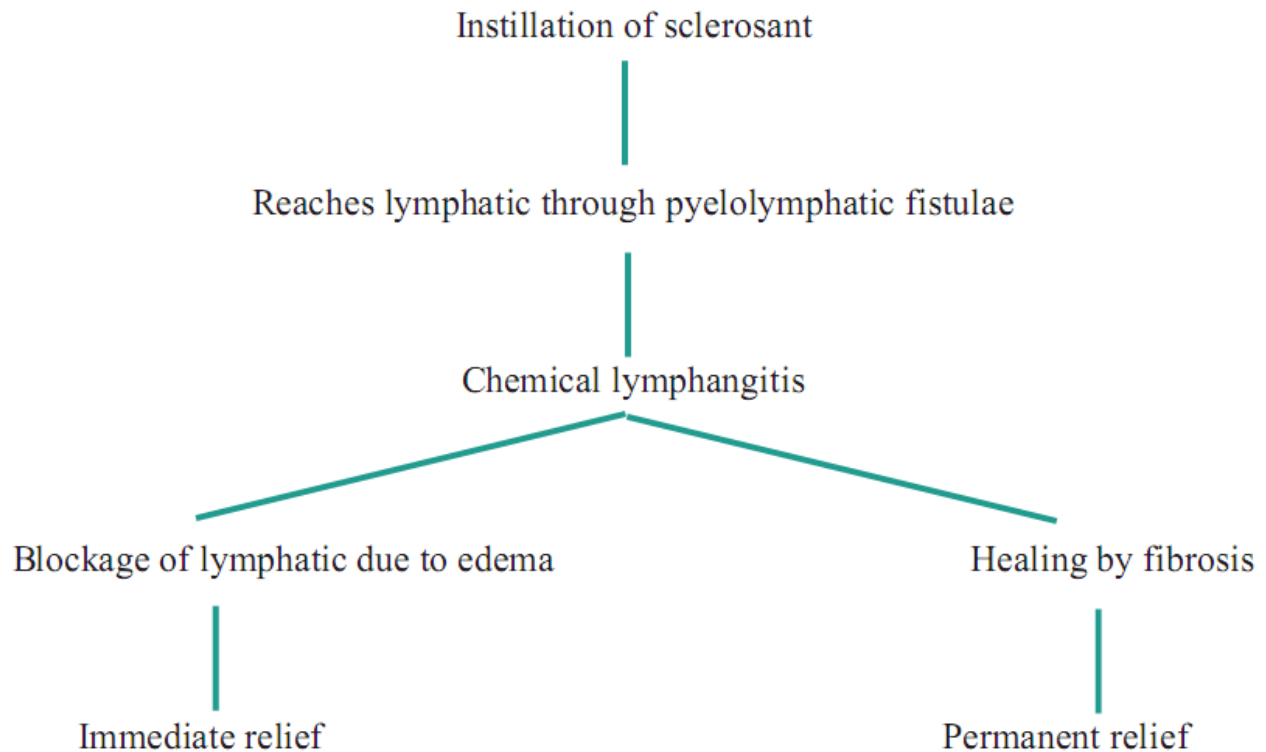
Abdominal binders may be applied during acute attacks of chyluria to reduce loss of chyle.

Cytoscopic bladder wash is recommended in cases of clot/chylous urine retention and/or recurrent urinary tract infections.

- **2) Sclerotherapy** := instillation of renal pelvis with chemicals - silver nitrate (0.1-3.0%), 0.2% povidone iodine, 15-25% sodium iodide, 10-25% potassium iodide, 50% dextrose, 76% hypertonic saline, combination therapy using 5/6 F ureteric catheter after localizing site on cystoscopy - in cases of failure of conservative management .
- These agents bring about relief by causing *chemical lymphangitis*

- Permanent relief: The procedure is performed under LA/ sedation with aseptic precaution keeping the head down and instilling 7- 10ml sclerosant (single dose) and/or albendazole 400mg may be given along with DEC as symptomatic treatment where filarial infection seems to be the cause.
 - Analgesic /antibiotic cover is given for at least 5 days.
 - **Dosage schedule** = 8 hourly for 3 days or 12 hourly for 2 days or weekly for 6-8 weeks.
 - Recurrence after 1st course can be treated for second time with high success rate.
 - In bilateral efflux= *gap* period of *minimum 6-8* weeks between 2 sittings is recommended .

n 1. Sclerotherapy – Mechanism of Action



- **Silver Nitrate (0.1-3.0%):** = **most** commonly used sclerosant till date. 2gm AgNO₃ powder is dissolved in 200ml of water in a bottle (black paper/dark room).

The need for sterilization (autoclaving) results in a 8-24 hr undesirable delay.

Other disadvantages =

water insolubility, susceptibility to light, and precipitation with normal saline to form insoluble silver chloride salts that may cause uretric obstruction. mortality has been reported due to acute tubular necrosis (**ATN**) on instillation of silver nitrate in both ureters simultaneously,

- **Povidone Iodine (0.2%):** It is a water soluble, non-ionic surfactant polymer that releases iodine slowly.

Prepared by mixing 2ml 5% povidone iodine to 8ml distilled water it is to be used as a fresh solution.

- **3) Heat-clearing and hemostatic treatment:** 26 out of 30 patients of chyluria have been reported to be cured completely with a basic heat-clearing and hemostatic prescription .
- **4) Surgical management:** treatment of choice in severe forms of chyluria

Indication= significant weight loss; hypoproteinemia, anasarca, and/or severe anemia;

recurrent clot retention and hematochyluria; recurrent UTI; refractory chyluria= (failure of conservative treatment with adequate dietary modification, medical management and two or more instillations of sclerosants);

clinically significant chyluria =(associated with chylous clot or hematochyluria or duration of more than 1 year or failure to respond to conservative medical measures) ;

altered immune status;

marked psychological disturbance.

- **Open surgical techniques=**

Lympho-venous disconnection: First described by Katamine in 1952 it is the most commonly performed procedure.

A fatty diet 24-36 hr prior to surgery facilitates detection of lymphatics. Pedal lymphangiography and/or methylene blue injection delineates the lymph channels. Reno-lymphatic disconnection operation is performed via a lumbar incision.

The kidney is freed, lymphatics in hilar area coursing along the renal vein are dissected, ligated & cut. Areolar tissue containing dilated lymphatics

travelling to the kidney in the perirenal and hilar

region are dissected and divided between ligatures thus stripping these structures completely. Hilar stripping is continued up to proximal 3-4cm ureter, the site for majority of shunts. Recurrence or bladder leak on imaging, mandates stripping of the entire ureter.

- **Patna operation (1977):** modified procedure of peri-ureteric lymphovenous stripping. Disconnection of only abdominal ureter is done without hilar disconnection. "Ureter in Lymphatic Tunnel" i.e. pampniform plexus in front and pre/para aortic plexus behind is the basic concept followed .
- **Omental wrapping (2004):** Omentum is used to wrap the renal vessels after stripping both anteriorly and posteriorly. This reduces the incidence of postoperative lymphatic drainage, lymphocele formation, recurrence, fibrosis and postoperative adhesions .
- **Renal autotransplantation:** rarely indicated for fistulas in the lower portion of ureter /bladder or when renal pedicle

stripping has failed (incomplete procedure/ recanalization) or there is formation of newer fistulas .

- ***Nephrectomy:*** It is used only for kidneys rendered non-functional following lymphovenous disconnection and serves as a life saving measure in severely refractory chyluria .

- ***Cockett and Goodwin procedure*** (1962):

based on diversion of a single hilar lymphatic to a spermatic/gonadal vein in end-to-side fashion.

It provides a safety valve mechanism for renal lymphatic-hypertension=the basis of chyluria.

It obviates need for complete stripping and there is a decreased incidence of lymphocutaneous fistula.

Demerits = increased operative time, technical difficulty and need for magnification .

- ***5) Microsurgical procedures:*** Recommended in the old and debilitated pt
- this technique useful in persistent or recurrent chyluria.
- Aiming= to decrease the intra lymphatic pressure by increasing lymph drainage into venous system, it reduces incidence of lymphangiectasia and facilitates healing of lymphatic fistula in the renal papilla.
- A simple, easy relatively superficial procedure , less traumatic to surrounding tissues

- it obviates the use of CPE / lymphangiography as there is no correlation

between site of operation or side of lympho-renal fistula.

Technically demanding - magnification is very essential as 90% of lymphatic vessels have diameter <1mm, making it difficult to finding vein of adequate diameter and in proper place (for anastomosis).

- **Lymphangiovenous anastomosis:** An end-to-end anastomosis is made in the inguinal region in men

and either in dorsum of foot/leg/thigh in women.

3 to 4 anastomoses per side are made using 4 to 6 interrupted (9-11) 0 nylon sutures ,

- **Lymph node – Saphenous vein anastomosis:**

An end-to-side anastomosis of inguinal lymph node to generally a tributary of saphenous vein (the distal end of which is ligated) is made. Generally a single anastomosis using 6 to 7 interrupted sutures is sufficient

- **Laparoscopic procedures:** Of the two main routes i.e. transperitoneal (peritoneum transgressed) and retroperitoneal (easy and straight access to kidney)

the latter approach significantly reduces incision – related morbidity without compromising the principles of open surgery .

- **Retroperitoneoscopy:** The technique comprise of nephrolympholysis, ureterolympholysis,

hilar vessel stripping, fasciectomy or nephropexy out of which the first three are mandatory while fasciectomy and nephropexy (to prevent renal pedicle torsion and nephroptosis) are not routinely recommended ,

added advantages of magnified view, better identification of lymphatics, better anastomatic results,

minimal morbidity, shorter hospital stay, excellent

- Retroperitoneoscopic renal pedicle lymphatic disconnection for chyluria completely ligates the lymphatic vessels and is a safe, effective and efficient surgical procedure with minimal invasion, less pain, less blood loss, lower morbidity, short hospital stay and rapid recovery . The procedure is recommended for treatment of recalcitrant chyluria .

- **6) Robotics:**

Nephrolympholysis, ureterolympholysis, hilar vessel stripping, fasciectomy and nephropexy can all be done as in conventional laparoscopy.

This state-of-the-art Master-Slave system provides 3-dimensional vision, seven degrees of freedom, 540 wristed movements, elimination of hand tremors, accurate scaling of

movements and ergonomic comfort in addition to the

advantages and objectives of open surgery.

- 7) Other therapies
- **Medical therapy:** Posttraumatic chyluria due to lymphorenal fistula regress after somatostatin therapy .

Chyluria has been reported to disappear following percutaneous sclerotherapy to the left inguinal lymph nodes with *doxycycline in a postoperative case of lymphangiomatosis* .

- Chyluria (after radical nephrectomy) = treated with *N-butyl-2-cyanoacrylate*
- **Gerota's fasciectomy** -bilateral excision of perinephric fascia and fat (Gerota's fasciectomy) in 2 stages has been reported to result in complete resolution of chyluria .
- **Endoscopic therapy:** Chyluria has been successfully treated in 5 patients with endoscopic coagulation using guide tube methods

RECURRENCE:

- Incomplete stripping, recanalization, reflux from contralateral side, reflux from terminal ureter or bladder are important causes of recurrence .

Diagnosed by radio imaging especially

lymphangioscintigraphy, lymphangiography (detects leaks at bladder level) and cystoscopy (blebs under mucosa / loosely hanging chylous clots from bladder wall).

This can however be prevented by use of magnifying loops, operating microscope, omental wrap technique or the use of laparoscope

COMPLICATION:

- **Complications=**
- After Sclerotherapy =anuria with pelvi-calyceal cast formation, acute necrotizing ureteritis and fatal renal and hepatic failure .
- Complications of retroperitoneoscopic surgery are few - inferior vena cava injury, renal segmental artery injury, lymphatic leak through the drain, and delayed wound healing.

Advancements in laparoscopy with the advent of robotics is a step forward to minimize the complication rate .