1/ Hello #Medtwitter #NephTwitter.
Different urine colors and the pathology it signifies is amazing!
Milky white urine is one such color we dread. What does it signify? Lets read on 📚
#ASPNFOAMgroup @ASPNeph #ASPNeph #pedneph #chyluria
2/
Let's see where this comes from....
"Chyle" is milky white lymphatic fluid composed of chylomicrons, comes from Greek word khÙlos or “juice”
Chyluria or milky urine is due to urinary-lymphatic communication

What the common causes of chyluria?
3/ All of the above
Filariasis, a parasitic disease caused by Wuchereria bancrofti, most commonly seen in adults and rarely reported in children
PMID:29549133.
Check out #NephMadness 2022 parasite region for description of filariasis

https://ajkdblog.org/2022/03/01/nephmadness-2022-parasites-region/#Filariasis

4/ Common causes of chyluria, depends on geographical location. Parasitic disease more common in Indian sub-continent and Sub-Saharan Africa

<table>
<thead>
<tr>
<th>Parasitic</th>
<th>Non-Parasitic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wucheria bancrofti (95%)</td>
<td>Trauma</td>
</tr>
<tr>
<td>Taenia</td>
<td>Congenital lymphatic malformation e.g.,</td>
</tr>
<tr>
<td></td>
<td>lymphatic-urinary fistula</td>
</tr>
<tr>
<td></td>
<td>Noonan, Turner, Gorham-Stout syndrome</td>
</tr>
<tr>
<td>Malaria</td>
<td>Post-surgery e.g. nephrectomy</td>
</tr>
<tr>
<td>Ankylostomiasis</td>
<td>Non-parasitic infections</td>
</tr>
<tr>
<td></td>
<td>Congenital stenosis of thoracic duct</td>
</tr>
<tr>
<td></td>
<td>Malignancy e.g., AML, testicular cancer</td>
</tr>
</tbody>
</table>

@nefron1310
How do the lymphatics work?
- Lymphatics reabsorb fluid and protein from tissues and extravascular spaces.
- No basement membrane so high permeability for reabsorption of proteins too.
- Located in deep dermis, parallel to veins, one-way flow.

Lymphatic system comprises of following:
- Afferent lymphatics
- Regional lymph nodes
- Efferent lymphatics
- Cisterna chyli
- Thoracic duct
- Subclavian vein
The Lymphatic System

- Cervical lymph nodes
- Thymus
- Palatine tonsil
- Axillary lymph nodes
- Spleen
- Thoracic duct
- Cisterna chyli
- Inguinal lymph nodes
- Right lymphatic duct
7/
Chyle composed of albumin+fibrin+emulsified fat
Non-parasitic chyluria is due to obstruction of normal lymphatic flow between intestines and thoracic duct, stasis, retrograde flow of lymph into renin pedicle lymphatic, lymphorenal fistula formation

8/
The regurgitation theory is based on rupture of ectatic lymphatics secondary to inflammation or toxic metabolite and releasing chyle into the renal system

PMID: 32728391
https://www.indianjurol.com/article.asp?issn=0970-1591;year=2005;volume=21;issue=1;spage=18;epage=23;aulast=Dalela

What condition mimics lymphatic filariasis in children?

9/
Yes! Nephrotic syndrome has anasarca, proteinuria, lets see how to differentiate by urine test.
This tweetorial focuses on non-parasitic causes of chyluria
Chyluria in children is rare and underreported; described mostly in case reports
PMID: 29549133
Clinical presentation in ~2/3 cases: milky white urine

- Symptoms of urinary infection
- Chyle colic
- Systemic symptoms: weight loss, chills, edema
- Chyluria increases with fatty-meal and decreases with fat-free diet or fasting (PMID: 20872078)

Examination of post prandial urine microscopy separates into fat, fibrin and cellular debris

Ether (fat solvent) or chloroform clears up urine

PMID 32728391

Image courtesy #ASPN Radiology Webinar
What is considered confirmatory test for chyluria on UA?

12/
- Urine TG $> 15$ is considered diagnostic

How to differentiate chyluria by urine tests?

- Milky/turbid urine
  - Phosphate: Clears up with 5% Acetic acid
  - Chyluria:
    - Clears up with Ether/chloroform
    - Sudan III test red-staining Fatty globules
    - + lymphocytes by Methylene blue test
    - Urine TG $> 15$ mg/dl
  - Pyuria:
    - + neutrophils

@nefron1310
Imaging

USG overall assessment of kidney and bladder
IVP not helpful as intra-renal or ureteric pressure is required to visualize the dilated para-calyceal lymphatics
CT scan identify fat within bladder and dilated lymphatics and more so after lymphangiogram

MR lymphangiogram is now widely described technique
+/- gadolinium
useful for pre-op assessment for reconstructive repairs
safe, has good spatial resolution

#ASPN Radiology Webinar
Retrograde pyelogram—fistulous connection between urinary collection system and lymphatic system
Endoscopic visualization in some cases when conservative management fails
Cystoscopic view on milky urine effluxing left ureteral orifice

PMID 19528993, 19528993
Fig. 2. Retrograde pyelogram demonstrating lymphatico-urinary communication on the right kidney. Note the extravasated lymphangiographic contrast material in the right renal calices (arrows).
Lymphangioscintigram Tc99 labeled study for road-mapping the lymphatic drainage and diagnosing fistula;
safe and non-invasive but lacks sufficient spatial resolution, involves radiation

https://tech.snmjournals.org/content/46/2/123
Lymphangiogram
- Site, size, and the number of fistulous communications sensitivity of 90%
- Dysplasia of the lymphatic vessels, beading of the thoracic duct, dilated cisterna chylii
- Invasive, technically challenging, time consuming

PMID 20675319
CT lymphangiography:

- conventional lymphangiogram followed CT imaging within 60 min
- enhanced identification of lymphatic anomalies and the distribution of collateral lymphatic vessel

PMID: 29470154
Conservative approach

Fat-free formula for babies/children on enteral formula

High protein diet, Medium Chain Triglycerides (MCT) oil, coconut oil as these are absorbed by portal system and not intestinal lymphatics

Vitamins ADKE

TPN in intractable cases
20/
Minimally invasive

- Sclerotherapy with 1% silver nitrate or 0.2% povidone iodine instillation into renal pelvis to close the fistulous connection
- Endoscopic fulguration of fistula opening

21/

- **Chylo-lymphatic disconnection**
  
  PMID: 9439397

- Autotransplantation

Nephrectomy if non-functional kidney

🔥 Complications - fibrosis, adhesions, lymphocele, recurrence, prolonged recovery, wound infection, injury to blood vessel

🎯 Only if minimally invasive procedures fail

22/

🎯 **Role of sirolimus in pure lymphatic malformation is shown to be beneficial**

https://jamanetwork.com/journals/jamadermatology/article-abstract/2784291


That's all folks, hope this tweetorial helps to work up the next case of milky urine you see! #Chyluria #urinalysis #Pisseprophets
Take Home Points

- Etiology differs based on geographical location
- Some cool urine tests helps differentiate chyluria from other causes of milky urine
- MR lymphography and CT lymphangiogram are helpful diagnostic tests
- Treatment with low fat diet, minimally invasive procedure, adjuvant sirolimus. Reconstructive surgery in refractory cases

For a case-based clinical discussion with a radiology expert login to @ASPNeph website, February webinar #Membereducation #ASPNFOAM group
Special thanks to @drM_sudha @md_abdulqader83
Until next time
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