Ashley Rawson

@Ashley_Rawson

23 Tweets • 2021-08-01 00:53:44 UTC • See on Twitter

rattibha.com ♥

1/

📢Hello #MedTwitter.

This month's @ASPNeph Renal Pathology webinar is all about #IgAVasculitis #IgAVN formerly known as #HSP.

Urine for a treat!

#nephtwitter

Let's do a quick poll! Iga Vasculitis is most common in which age group?

2/

A: <10 years, about 90% of cases of IgA Vasculitis are seen in children under 10

🤯


YES!

3/

Let's talk numbers-

1️⃣ Kidney involvement in 30-50%

2️⃣ Kidney involvement appears within 6 weeks in 91%

3️⃣ IgA Vasculitis preceded by URI in 50%

4️⃣ If nephro findings, 10% progress to ESRD/severe CKD in 10 years

PMID: 139081 PMID: 32356414 PMID: 32060820

4/
What's with the two names? PMID: 32356414

HSP 👉 Johann Schönlein 👨‍🔬 and Eduard Henoch 👨‍🎓 made the first detailed analysis of this disease.

IgA Vasculitis 👉 Based on IgA deposited in blood vessel walls.

Check this article out on the two terms:

https://libgallay.com/2021/01/13/iga-vasculitis-terms/

5/
Tetrad of findings:
⚡Palpable purpura
⚡Joint pain and/or swelling
⚡GI issues- pain, intussusception, hematochezia
⚡Renal Involvement- hematuria, proteinuria, elevated creatinine
Uptodate

6/

How about another question?!

Can we prevent kidney disease associated with IgA vasculitis?? 😷🥼🏥

7/

A: Sadly, No...

😞

Evidence suggests that steroids early in IgA vasculitis don't prevent kidney involvement.

PMID: 26258874

8/

We still aren't sure why IgA vasculitis happens but we have some ideas... 💡

PMID: 23684700

Bacteria, viruses, and protozoa suggested as possible triggers:

Type of pathogen	Etiologic agent
Bacteria	Group A beta-hemolytic *Streptococcus pyogenes*
	Neisseria meningitidis
	Mycoplasma pneumoniae
	Bartonella henselae
	Helicobacter pylori
	Salmonella enteritidis
	Mycobacterium tuberculosis
	Staphylococcus aureus
	Chlamydophila pneumoniae
	Campylobacter jejuni
	Kingella kingae

	Kingella kingae
Viruses	Parvovirus B19
	Hepatitis B virus
	Hepatitis A virus
	Hepatitis C virus
	Varicella-zoster virus
Protozoa	*Giardia intestinalis*
	Trichomonas hominis
	Ascaris lumbricoides
	Entamoeba histolytica
	Plasmodium falciparum

9/

🧬 Some genes may predispose children:

PMID: 23684700

Gene	Gene name
HLA gene family	*HLA-DRB1*01*
	*HLA-DRB1*11*
	*HLA-DRB1*13*

	*HLA-A*26(*2601), HLA-B*35(*3503), HLA-B*52*
	*HLA-B*35* (*3503)
	HLA-A2, HLA-A11, HLA-B35
Genes involved in the synthesis of inflammatory and anti-inflammatory proteins	*MEFV*
	IL8 (polymorphic allele A in the interleukin 8 gene)
	IL8 (2767 A/G polymorphism in the interleukin 8 gene)
	IL1RN (polymorphic allele 2 [ILRN*2] in the interleukin 1 receptor antagonist gene)
	IL1 (– 511 C/T polymorphism in the interleukin 1-beta gene)
	TGF (TT genotype of the C-509T polymorphism in the transforming growth factor-beta promoter gene)
Genes involved in the regulation of endothelial function	*ACE* (D/I polymorphism in the intron 16 of the angiotensin converting enzyme gene)
	AGT (M235T mutation in the angiotensinogen gene)
	NOS2A (CCTTTn polymorphic

	alleles in the nitric oxide synthase promoter gene)
	VEGF (− 1154 G > A and − 634 G > C polymorphisms in the vascular endothelial growth factor gene)
Complement gene family	*C4B* (C4B*Q0 allele in the C4B gene)
Genes involved in the synthesis of antioxidant proteins	*PON1* (QQ genotype of the Q/R192 polymorphism in the paraoxonase 1 gene)

10/
What causes IgA Vasculitis??

Hastings et al. recently published an update on the pathogenesis of IgA vasculitis with nephritis. Check out their 4 hit hypothesis:
PMID: 33818625

Hit #1

Elevated circulatory Gd-IgA1

Hit #2

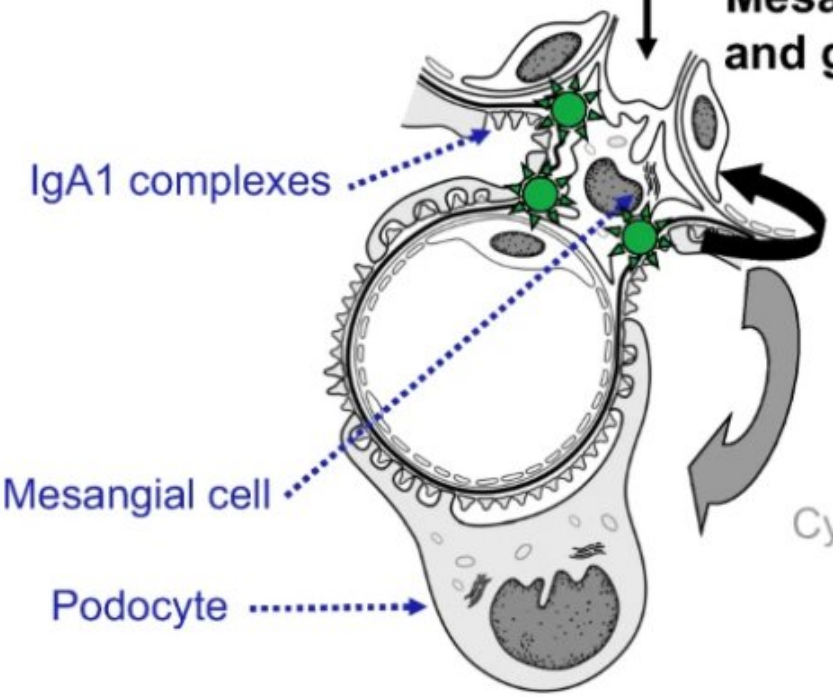
Production of IgG autoantibodies

Hit #3

Formation of IgG–Gd-IgA1-containing circulating immune complexes

Hit #4

Mesangial deposition and glomerular injury



11/

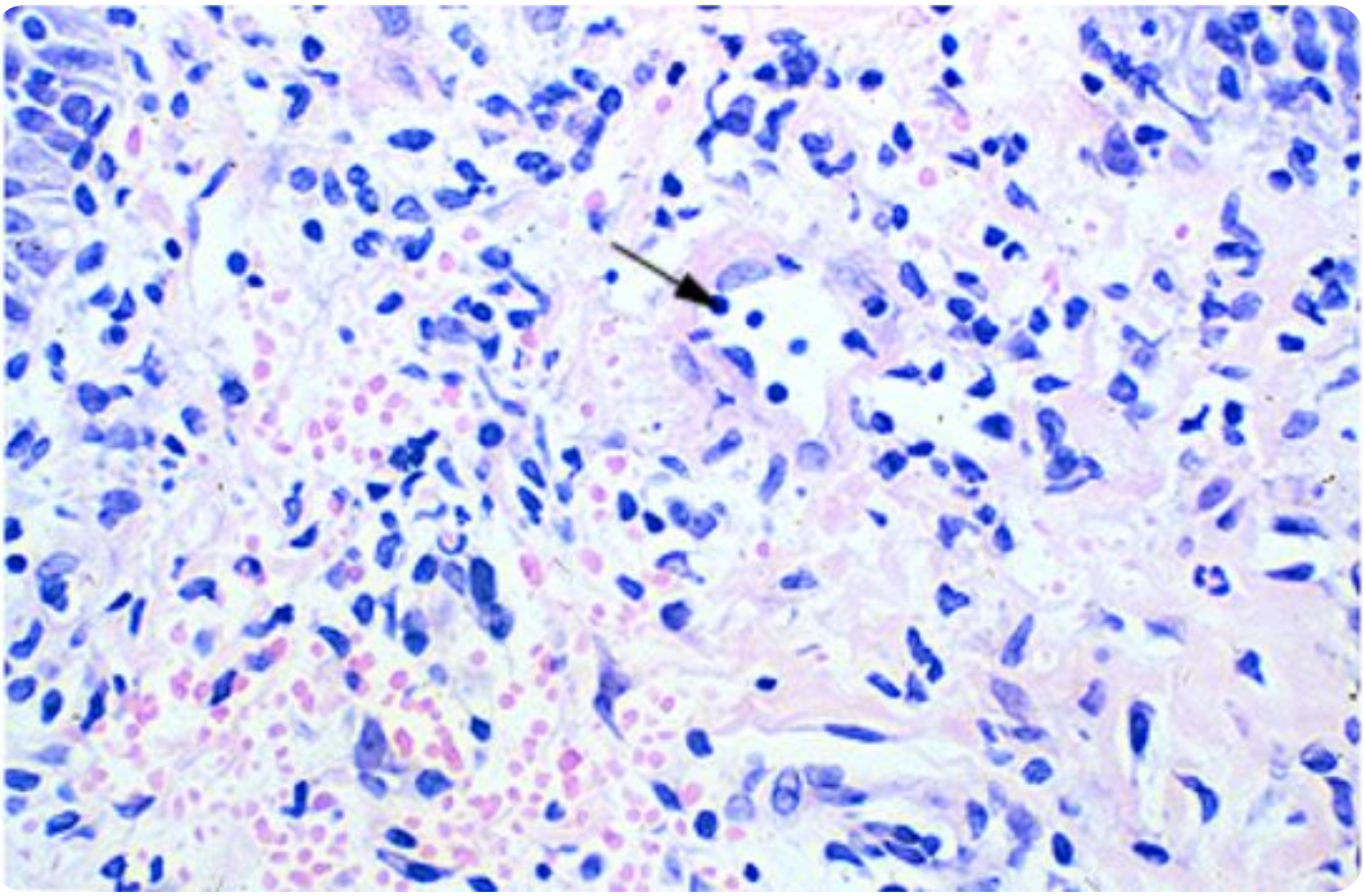
Diagnosis- usually clinical manifestations

🤦‍♀️ Not obvious? 🤷‍♀️ Not sure? 🤷‍♂️ Don't know?

A skin biopsy can help! 🦸‍♂️

PMID 9366584

Leukocytoclastic vasculitis in postcapillary venules with IgA deposition in small blood vessels of superficial dermis.



12/

What about a kidney biopsy? 🔬

Who gets one??

✅Nephrotic-range proteinuria

✅Elevated creatinine

13/

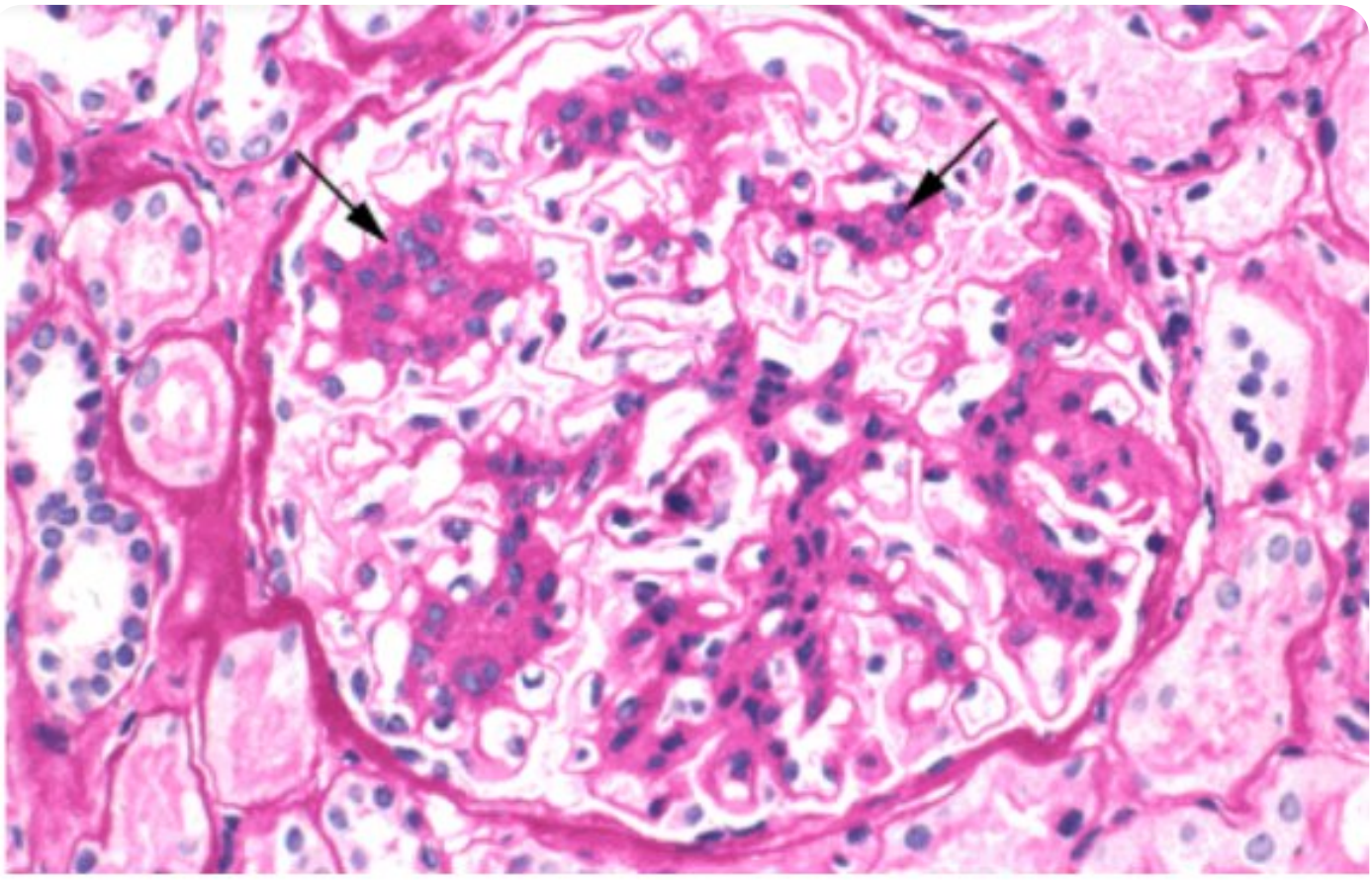
Kidney biopsy findings:

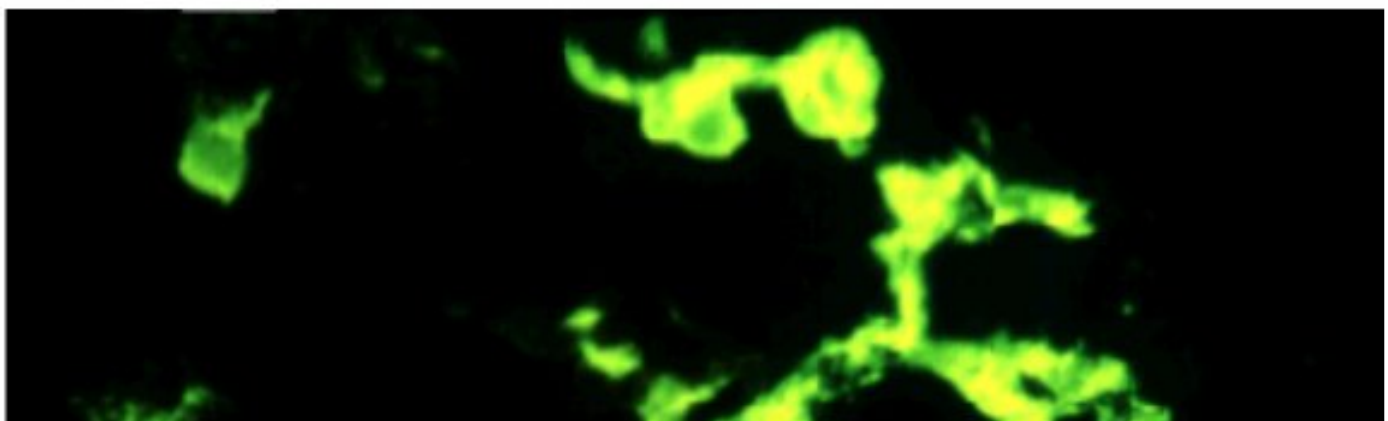
Uptodate

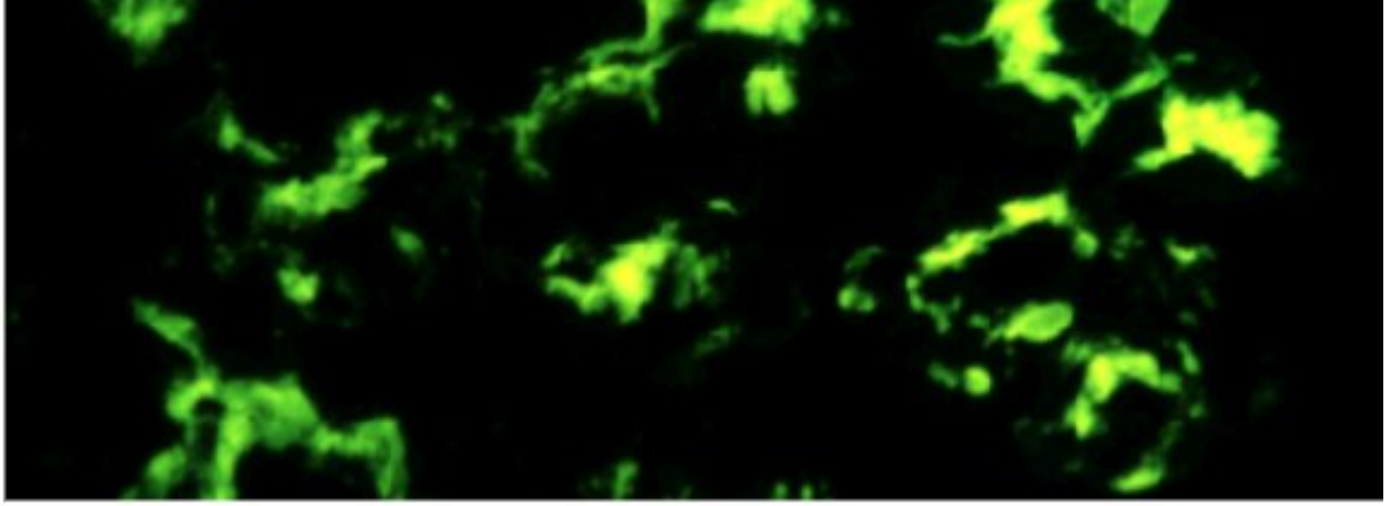
LM: mesangial proliferation ↔️ severe crescentic glomerulonephritis

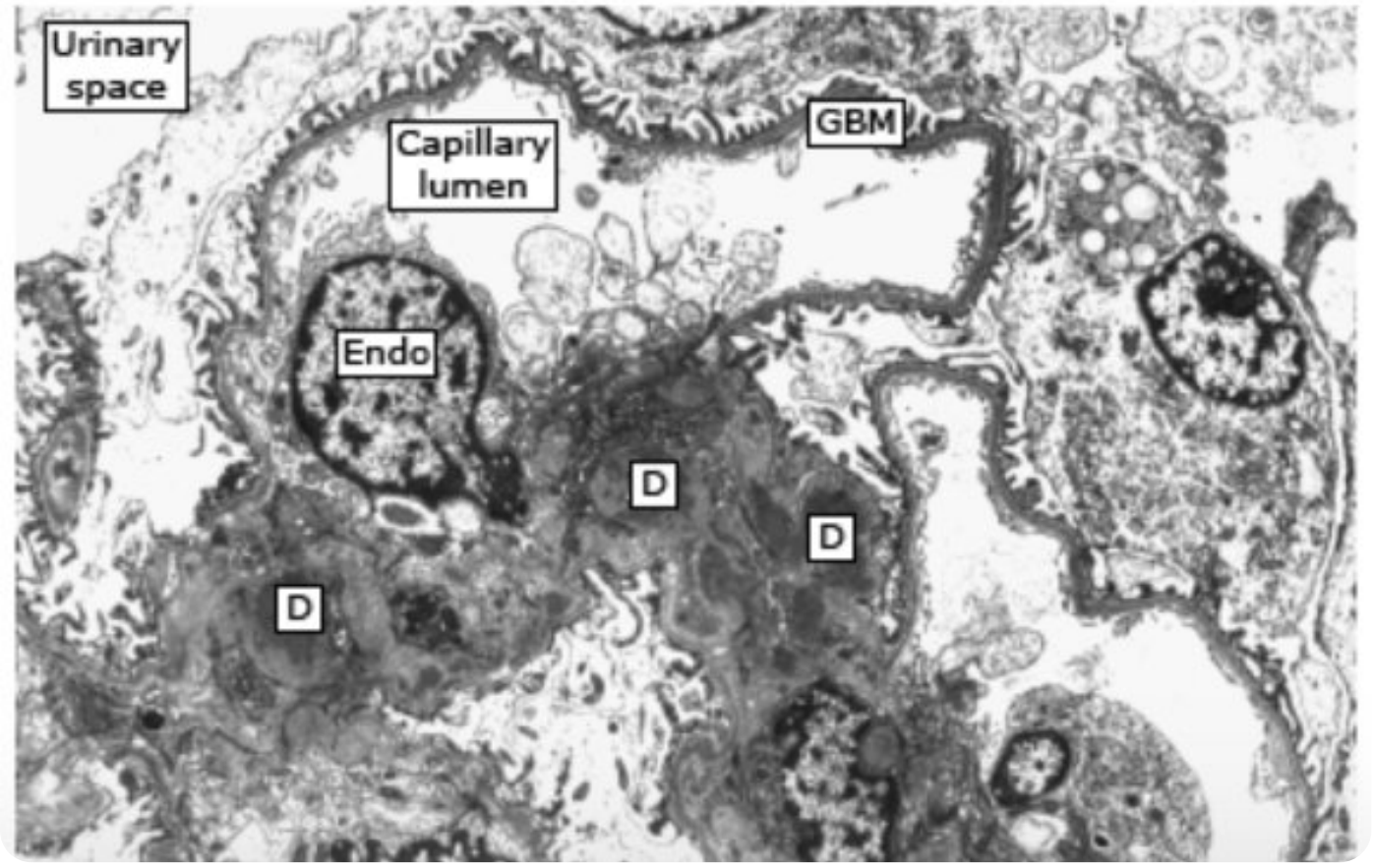
IF: IgA deposition in the mesangium

EM: electron-dense deposits in mesangial areas, occ extends into peripheral capillary loops









14/

The ISKDC classification has long been used to evaluate severity of kidney involvement on biopsy:

PMID: 28197887

🕵️📜

ISKDC grade	Description
Grade I	Minimal alterations
Grade II	Mesangial proliferation
Grade III	Proliferation or sclerosis with < 50% crescents ((a) focal or (b) diffuse)
Grade IV	Mesangial proliferation or sclerosis with 50–75%, crescents ((a) focal or (b) diffuse)
Grade V	Mesangial proliferation or sclerosis with > 75% crescents ((a) focal or (b) diffuse)
Grade VI	Membranoproliferative like glomerulonephritis

15/

The Oxford MEST-C classification used for IgAN is also being used more and more in IgA vasculitis: 🕵️📜

Histological feature	Score
Mesangial hypercellularity	M0: ≤50% of glomeruli M1: >50% of glomeruli
Endocapillary hypercellularity	E0: Absent E1: Present
Segmental glomerulosclerosis	S0: Absent S1: Present
Interstitial fibrosis and tubular atrophy	T0: 0–25% of cortical area T1: 26–50% of cortical area T2: >50% of cortical area
Cellular or fibrocellular crescent	C0: Absent C1: <25% of glomeruli C2: ≥25% of glomeruli

16/

What is reported on your biopsy reports?

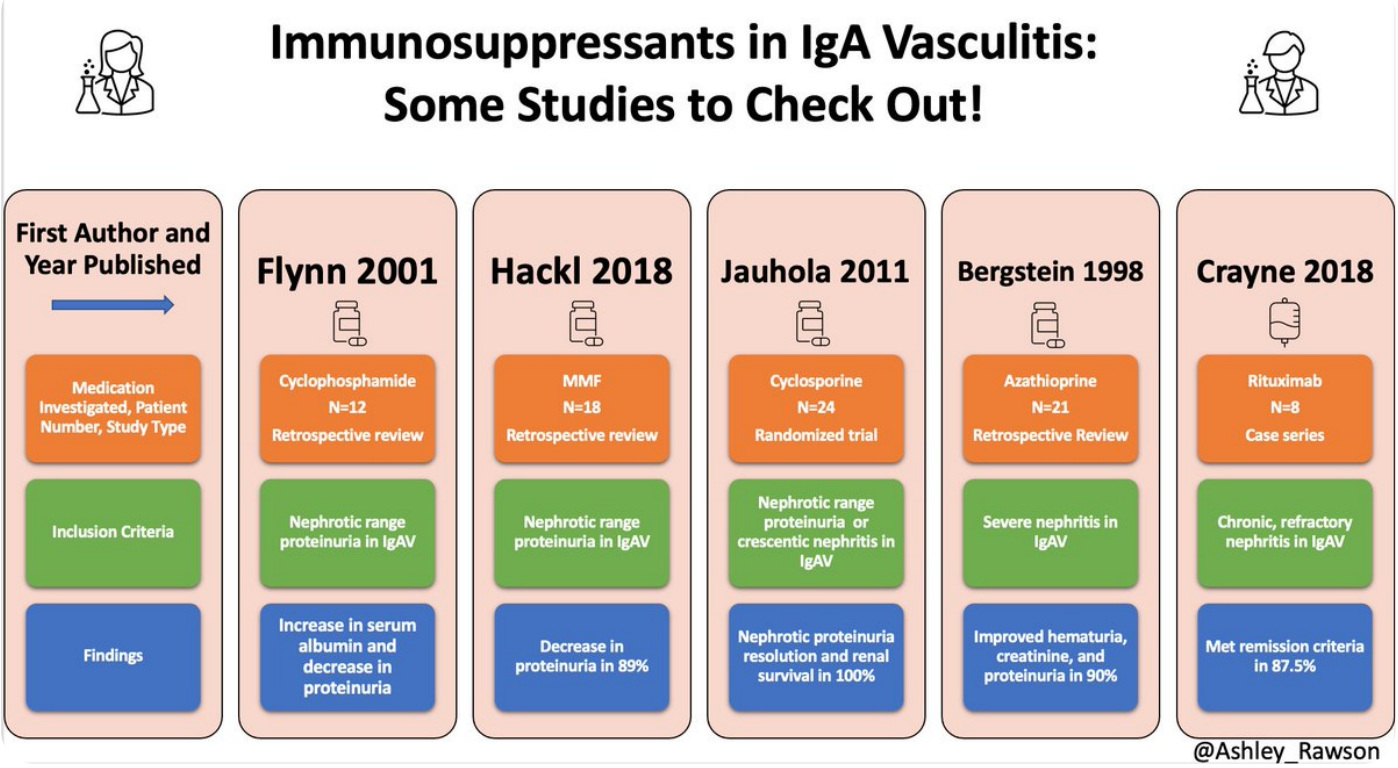
17/
Monitoring for kidney involvement in IgA vasculitis is very important. Children should have a urine dipstick performed weekly for 6 months (97% of children with IgA Vasculitis who develop renal involvement do so within 6 months, 1st morning void preferred).
Uptodate

18/
To finish things off:

How do we treat?💊💉🏥

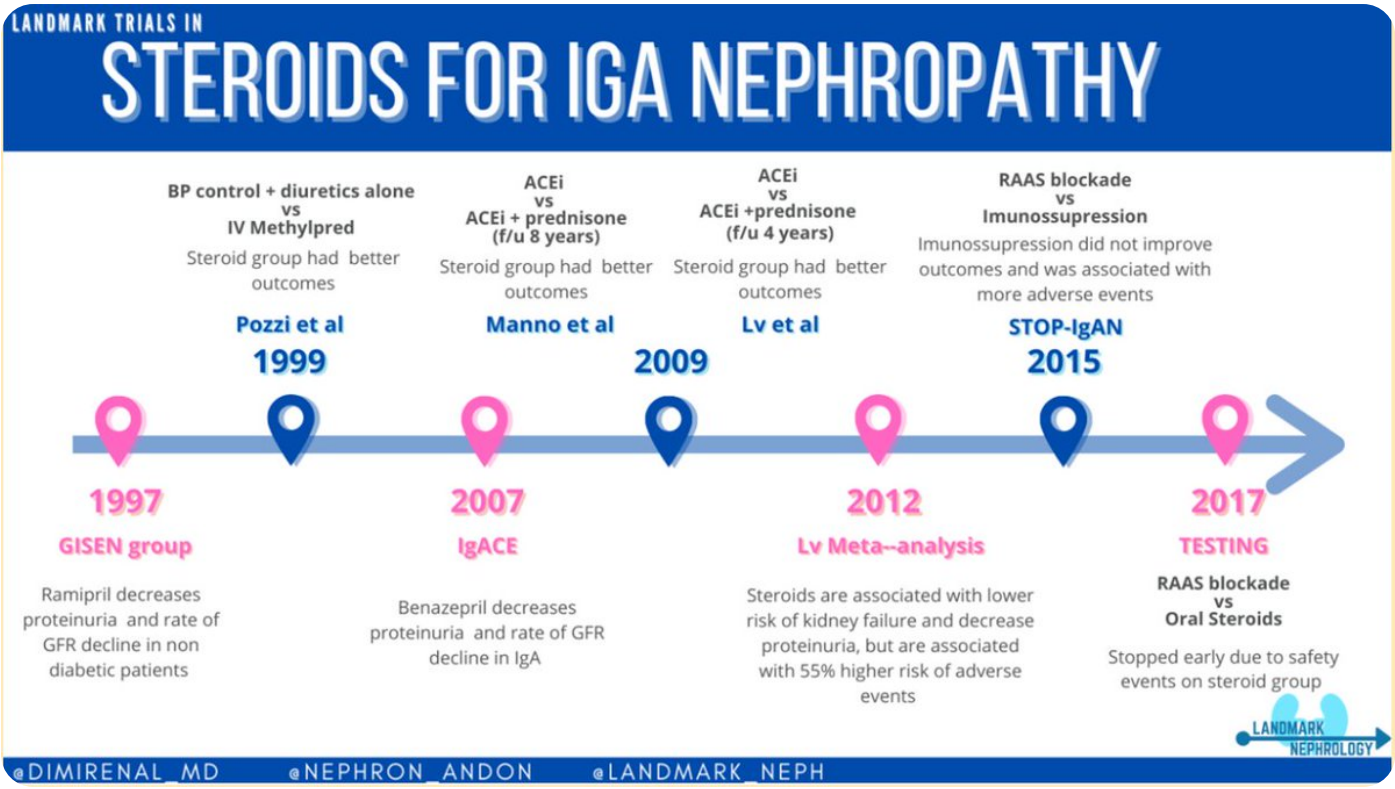
19/

Info on IgA vasculitis treatment is limited but here are some studies you can look into!



20/

@Landmark_neph, @nephron_andon, and @dimirenal_md also created a great timeline looking into IgA vasculitis and steroids.



21/

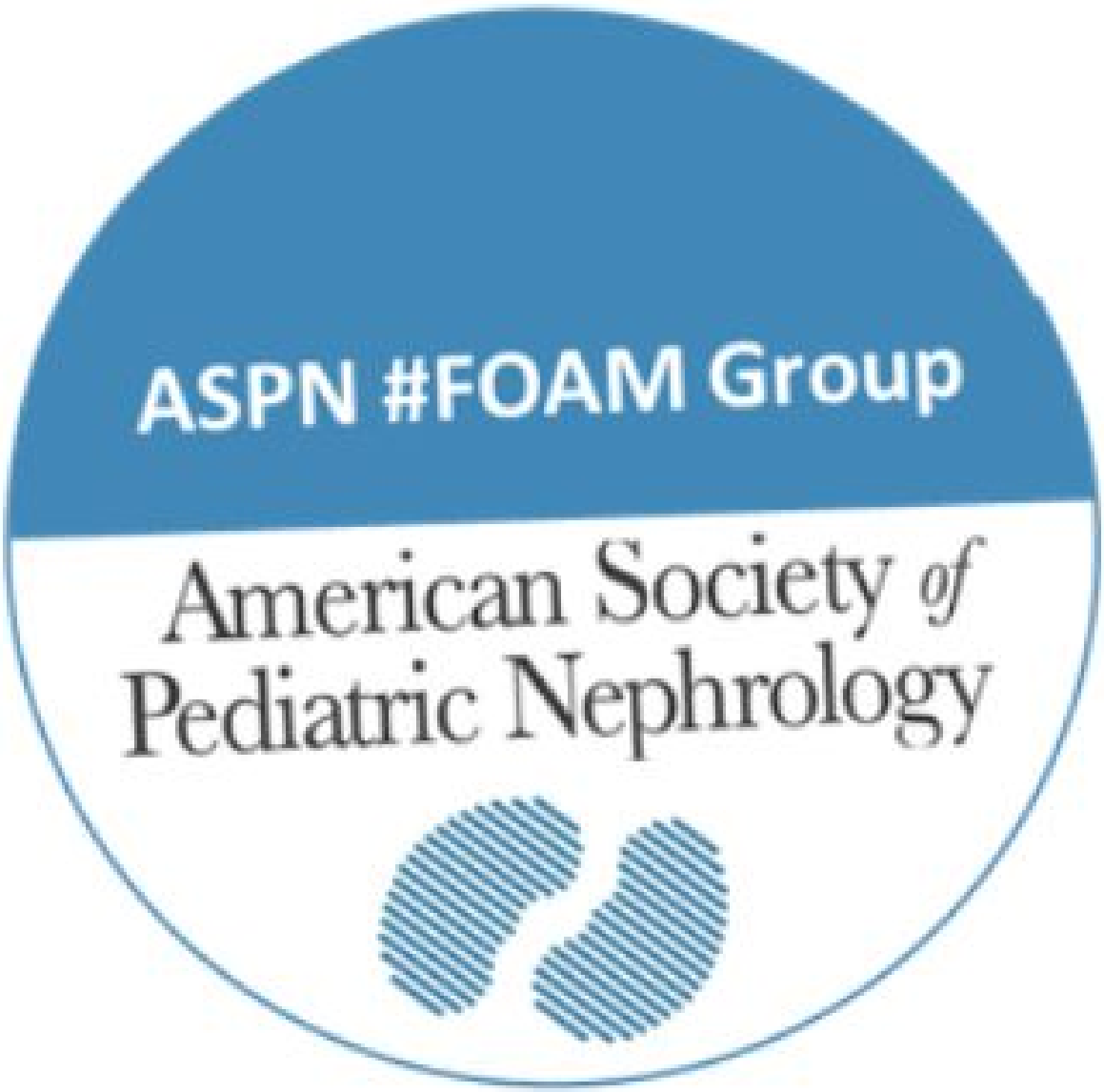
Refer to KDIGO 2012, European guidelines, and newer studies to get even more information on treatment.

Reply to this tweet and let us know your thoughts!

22/
Interested in learning more?? 🔎🔎

For a case-based discussion with pathologist and expert login to @ASPNeph website, July webinar. Answer questions to get #MOC2credits #Membereducation #ASPNFOAMgroup

@drM_sudha @menonshina @yardleyjojo @Trumidor
and Smitha Vidi



23/

Special thanks to @drM_sudha @priti899 @SwastiThinks

and @RoshanPGeorgeMD for all your help making my first tweetorial!

attachments and external links published (collectively referred to as "this publication"), were created at the request of a user (s) from Twitter. Rattibha provides an automated service, without human intervention, to copy the contents of tweets from Twitter and publish them in an article style, and create PDF pages that can be printed and shared, at the request of Twitter user (s). Please note that the views and all contents in this publication are those of the author and do not necessarily represent the views of Rattibha. Rattibha assumes no responsibility for any damage or breaches of any law resulting from the contents of this publication.