VASCULITIS POR IGA
(Purpura de Schonlein Henoch)

DRA CLAUDIA JUNCOS
NEFROLOGÍA PEDIÁTRICA
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CONSENSO DE CHAPEL HILL 2012

Small-vessel vasculitis (SVV)

A. Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV)

-Microscopic polyangiitis (MPA)

-Granulomatosis with polyangiitis (Wegener granulomatosis) [GPA (WG)]

-Eosinophilic granulomatosis with polyangiitis (Churg–Strauss syndrome) [EGPA (CSS)]

B. Immune complex SVV

-Anti-glomerular basement membrane (anti-GBM) disease

-Cryoglobulinemic vasculitis (CV)

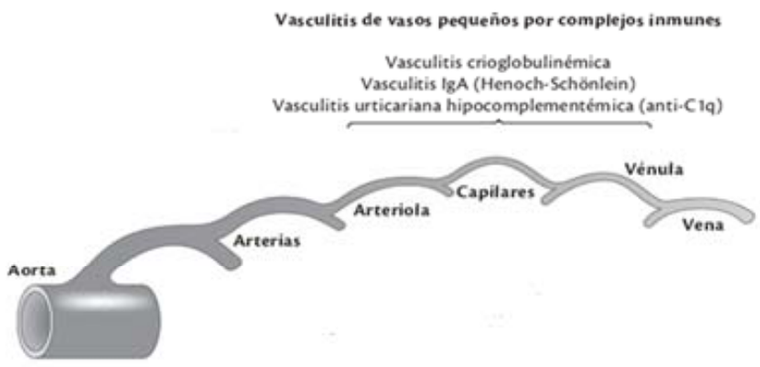
-IgA vasculitis (Henoch–Schönlein) [IgAV (HSP)]

-Hypocomplementemic urticarial vasculitis (HUV) (anti-C1q vasculitis)

Pediatr Nephrol (2015) 30:1425–1432
DOI 10.1007/s00467-014-3015-0

EDUCATIONAL REVIEW

Vasculitis: do we know more to classify better?



Adaptada de Jennette JC *et al.* 2012. Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides. Arthritis Rheum, 2013;65(1):1-11.

Criteria

EDITOR'S CHOICE

EULAR/PRINTO/PRES criteria for Henoch–Schönlein purpura, childhood polyarteritis nodosa, childhood Wegener granulomatosis and childhood Takayasu arteritis: Ankara 2008. Part II: Final classification criteria

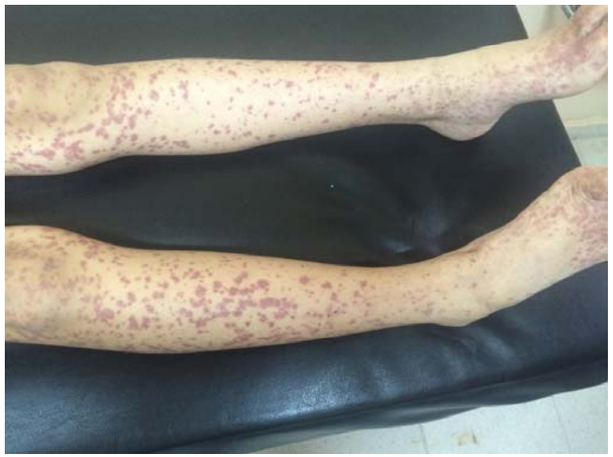
Seza Ozen,[1] Angela Pistorio,[2] Silvia M Iusan,[3] Aysin Bakkaloglu,[1] Troels Herlin,[4] Riva Brik,[5] Antonella Buoncompagni,[3] Calin Lazar,[6] Ilmay Bilge,[7] Yosef Uziel,[8] Donato Rigante,[9] Luca Cantarini,[10] Maria Odete Hilario,[11] Clovis A Silva,[12] Mauricio Alegria,[13] Ximena Norambuena,[14] Alexandre Belot,[15] Yackov Berkun,[16] Amparo Ibanez Estrella,[17] Alma Nunzia Olivieri,[18] Maria Giannina Alpigiani,[19] Ingrida Rumba,[20] Flavio Sztajnbok,[21] Lana Tambic-Bukovac,[22] Luciana Breda,[23] Sulaiman Al-Mayouf,[24] Dimitrina Mihaylova,[25] Vyacheslav Chasnyk,[26] Claudia Sengler,[27] Maria Klein-Gitelman,[28] Djamal Djeddi,[29] Laura Nuno,[30] Chris Pruunsild,[31] Jurgen Brunner,[32] Anuela Kondi,[3] Karaman Pagava,[33] Silvia Pederzoli,[3] Alberto Martini,[3,34] Nicolino Ruperto[3]; for the Paediatric Rheumatology International Trials Organisation (PRINTO)

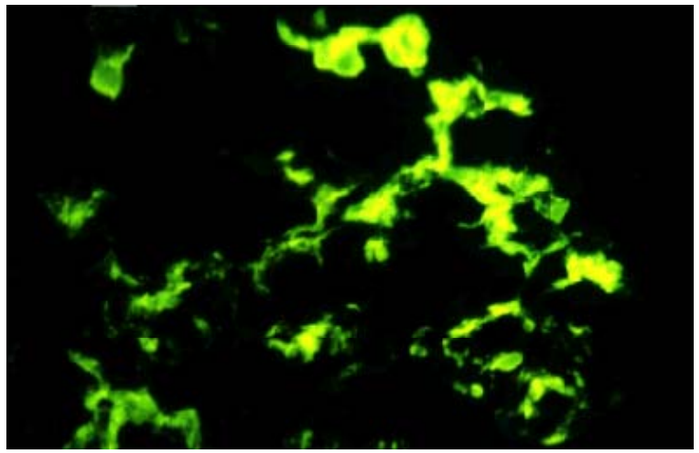
ANKARA 2008

Pediatr Nephrol (2015) 30:1425–1432

ACR criteria versus Ankara 2008 criteria regarding vasculitis classification

Vasculitis	Classification criteria [reference number]	
	ACR criteria	Ankara 2008 criteria
IgA vasculitis/HSP	≥2 of the following: -≤20 years of age at disease onset -Palpable purpura -Acute abdominal pain -Biopsy showing granulocytes in the wall of small arterioles/venules	Purpura or petechia (mandatory) with lower limb predominance plus 1 of 4: -Abdominal pain -Histopathology (predominant IgA deposit in a biopsy) -Arthritis or arthralgia -Renal involvement





VASCULITIS POR IGA

- Vasculitis sistémica mas común en niños
- 6 a 24/100.000 año
- 90% menores de 10 años. Media 6 años
- Compromiso renal en 30-50% de pacientes
- Pronóstico a largo plazo se relaciona con compromiso renal

Pediatr Nephrol
DOI 10.1007/s00467-014-2815-6

EDUCATIONAL REVIEW

Henoch–Schönlein purpura nephritis

Martin Pohl

Received: 8 December 2013 / Revised: 18 March 2014 / Accepted: 20 March 2014

REVIEWS

Davin, J.-C. & Coppo, R. *Nat. Rev. Nephrol*. **10**, 563–573 (2014); published online 29 July 2014

Henoch–Schönlein purpura nephritis in children

Jean-Claude Davin and Rosanna Coppo

KIDNEY DISEASE · IMPROVING GLOBAL OUTCOMES ®

KDIGO

2012

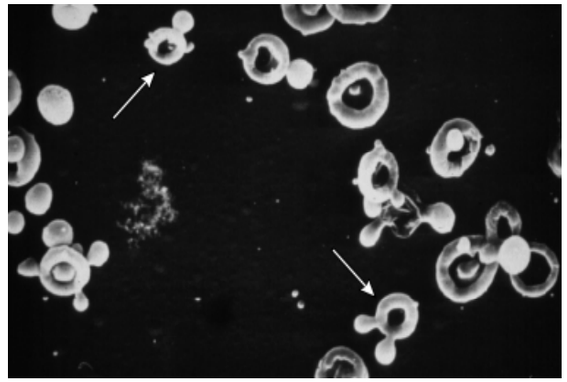
KDIGO 2012

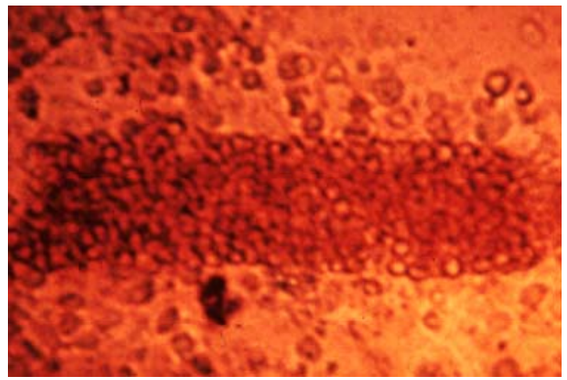
12 Estudios 1.133 Niños

34% Orina patológica

79% Hematuria con o sin proteinuria

21% Sindrome Nefrítico Sindrome Nefrótico





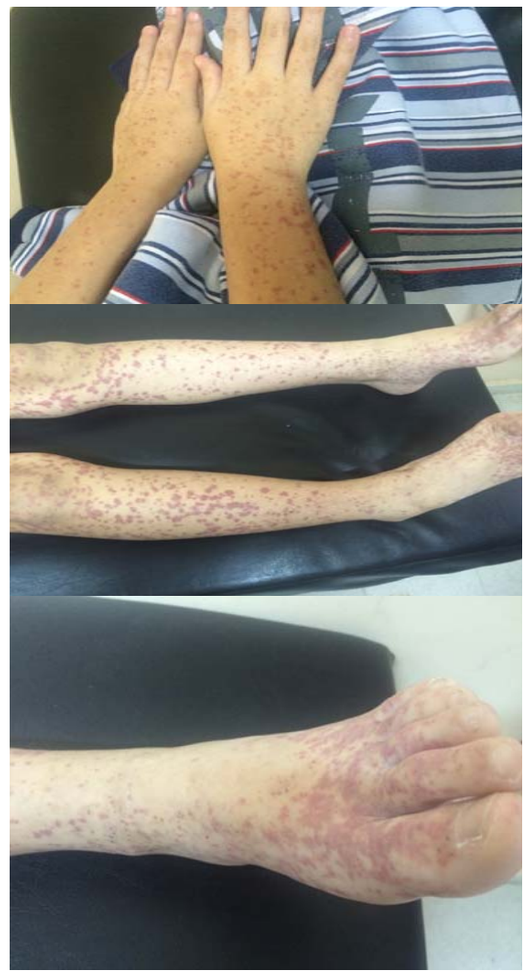
NEFRITIS POR VASCULITIS IgA

Asociada

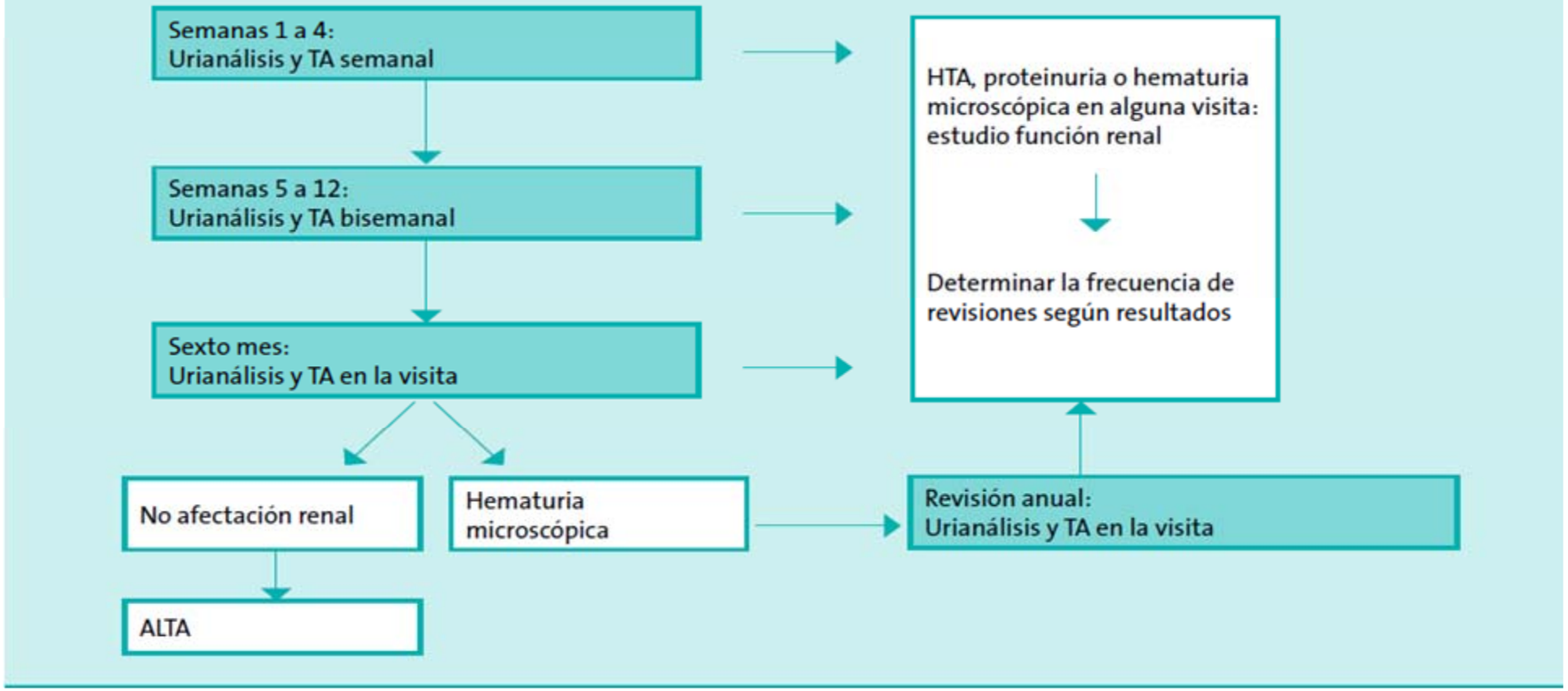
- Mayor edad a la pesentación
- Púrpura persistente
- Recurrencia de la púrpura
- Síntomas abdominales graves

Presentación

- 85% primeras 4 semanas
- 90% primeras 8 semanas
- 97% dentro de los 6 meses



Propuesta de seguimiento ambulatorio en los pacientes con púrpura de Schönlein-Henoch



PÚRPURA DE SCHÖNLEIN-HENOCH

S Ricart Campos
Unidad de Reumatología Pediátrica. Servicio de Pediatría. Hospital Sant Joan de Déu. Barcelona

Ricart Campos S. Púrpura de Schönlein-Henoch.
Protoc diagn ter pediatr. 2014;1:131-40

Sindrome Nefrótico/ Nefrítico / Fallo renal

↓

Alto riesgo de compromiso renal crónico

↓

Seguimiento prolongado

KIDNEY DISEASE · IMPROVING GLOBAL OUTCOMES ® KDIGO

ORIGINAL ARTICLE

Risk of long term renal impairment and duration of follow up recommended for Henoch-Schönlein purpura with normal or minimal urinary findings: a systematic review

H Narchi

Arch Dis Child 2005;90:916–920. doi: 10.1136/adc.2005.074641

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Pediatr Nephrol
DOI 10.1007/s00467-017-3794-1

ORIGINAL ARTICLE

Presentation of pediatric Henoch–Schönlein purpura nephritis changes with age and renal histology depends on biopsy timing

Received: 18 June 2017 / Revised: 2 August 2017 / Accepted: 20 August 2017

Presentation on diagnosis of HSP and long term renal impairment

	Children		Long term renal impairment		
	No.	%	No.	% (95% CI)	Relative risk (95%)
Total	1133		21	1.8 (1.1 to 2.8)	NA
With normal urine	746	65.8	0	0 (0 to 0.5)	NA
With abnormal urinalysis	387	34.2	21	5.4 (3.3 to 8.3)	NA
Isolated haematuria ± proteinuria	305	78.8	5	1.6 (0.5 to 3.8)	(baseline)
Nephritic or nephrotic syndrome	82	21.2	16	19.5 (11.1 to 31.7)	11.9 (4.1–41.5)

NA, not applicable; CI, confidence intervals.

Severidad Clínica Inicial

+

Hallazgos Histológicos

→

Pronóstico a Largo Plazo

Henoch-Schönlein Purpura Nephritis: Pathophysiology, Treatment, and Future Strategy

Jean-Claude Davin

Clin J Am Soc Nephrol 6: 679–689, 2011. doi: 10.2215/CJN.06710810

Pediatr Nephrol
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CrossMark

ORIGINAL ARTICLE

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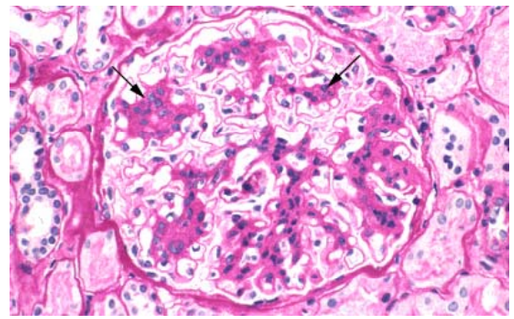
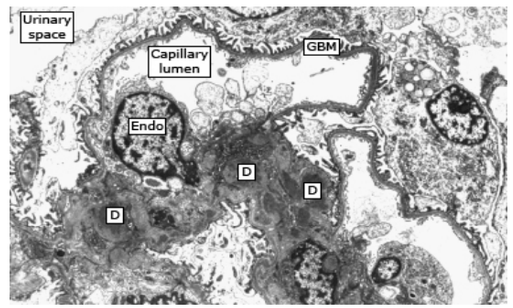
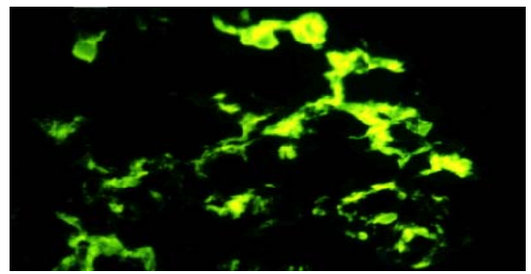
INDICACION DE BIOPSIA RENAL

- Sindrome Nefrítico
- Fallo Renal Agudo
- Sindrome Nefrótico
- Proteinuria mayor 1 gr/m^2 / día durante 4 a 6 semanas



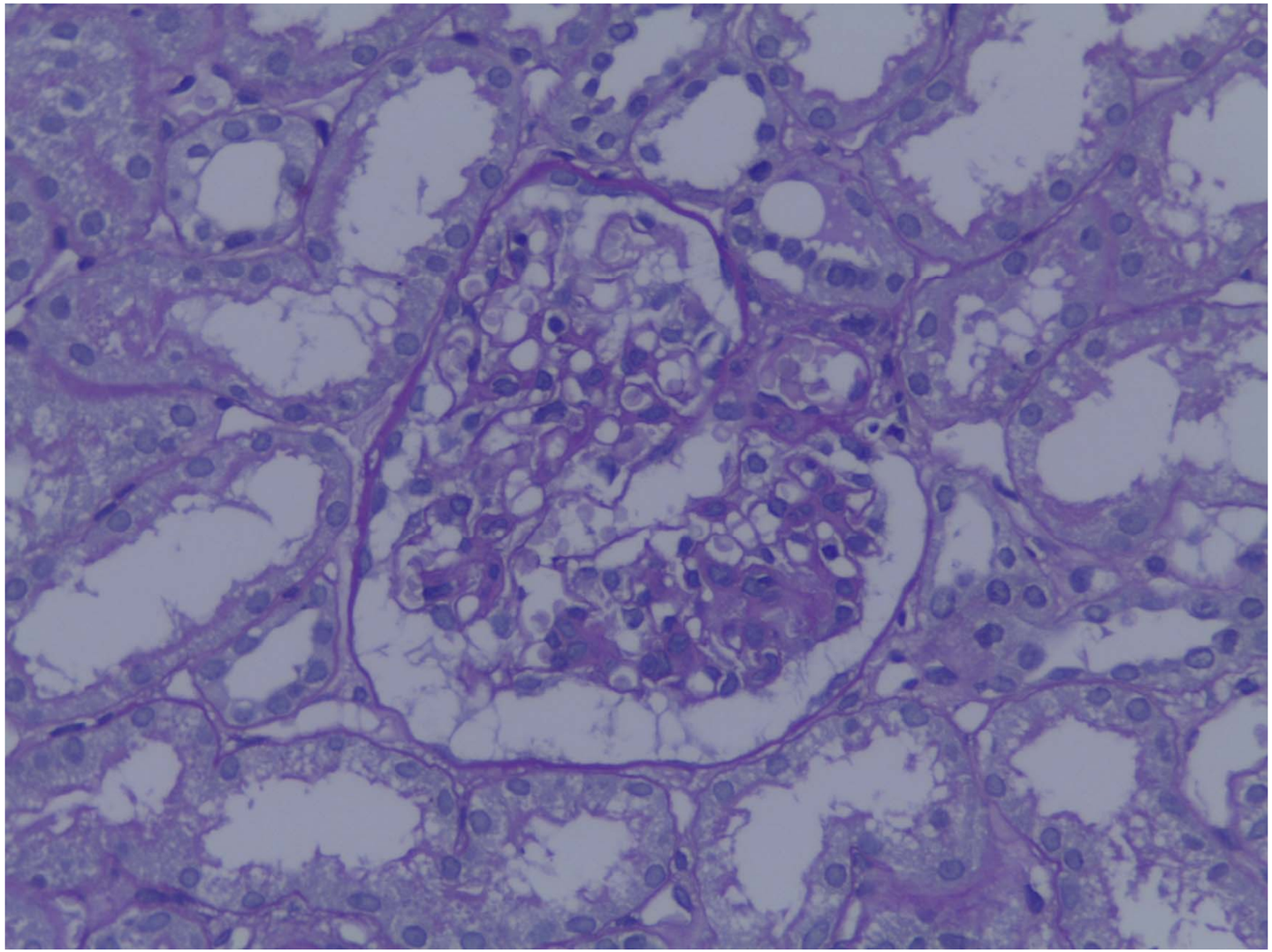




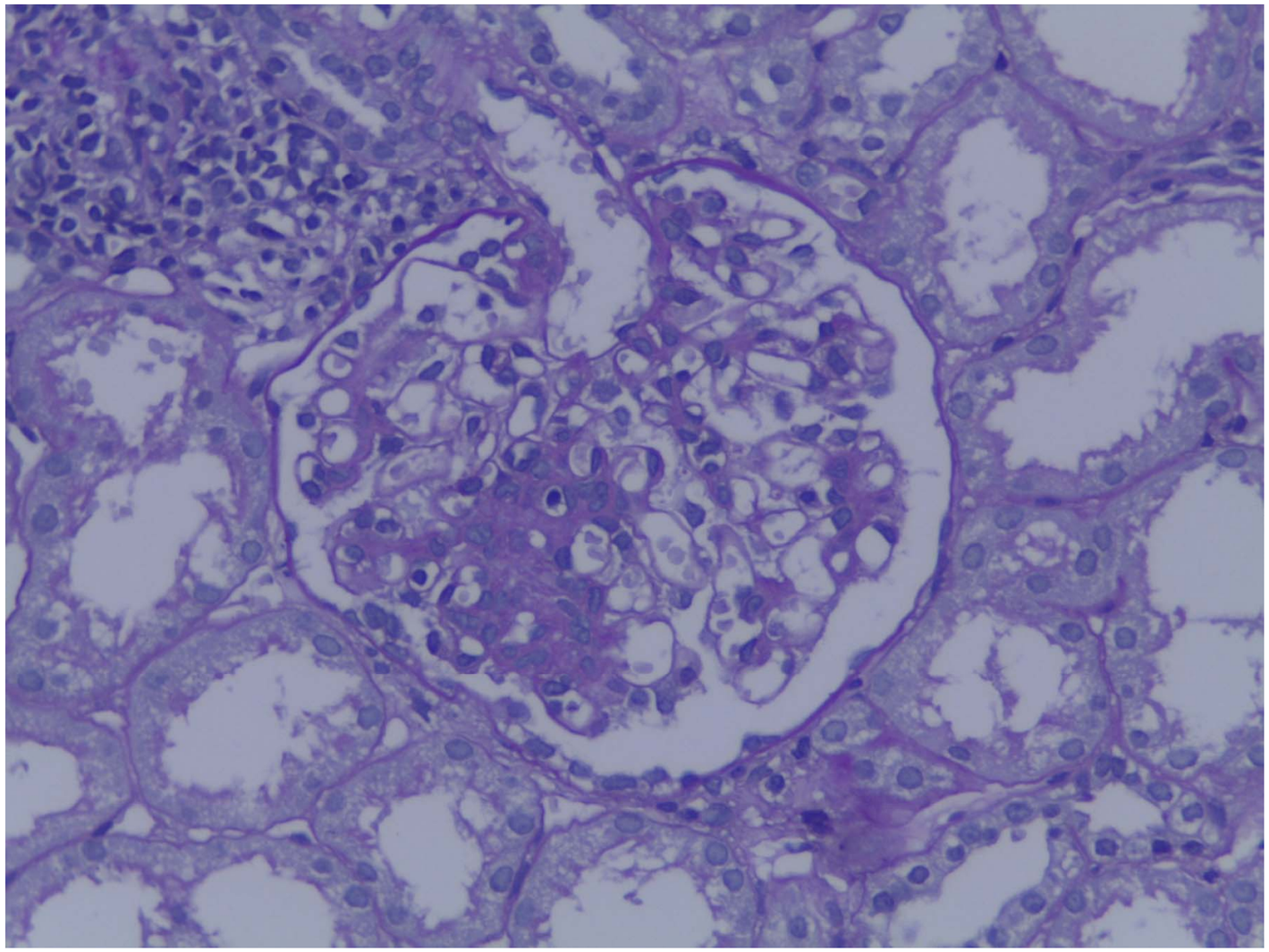
ISKDC

Grado ISKDC	
I	Alteraciones mínimas
II	Proliferación Mesangial
III A	Proliferación focal con < 50 % semilunas
III B	Proliferación difusa con < 50 % semilunas
IV A	Proliferación focal con 50 a 75 % semilunas
IV B	Proliferación difusa con 50 a 75 % semilunas
V A	Proliferación focal con > 75 % semilunas
V B	Proliferación difusa con > 75 % semilunas
VI	Glomerulonefritis Membranoproliferativa

ISKDC = International Study of Kidney Diseases in Children



MO Hospital Infantil Córdoba,
Diciembre 2017



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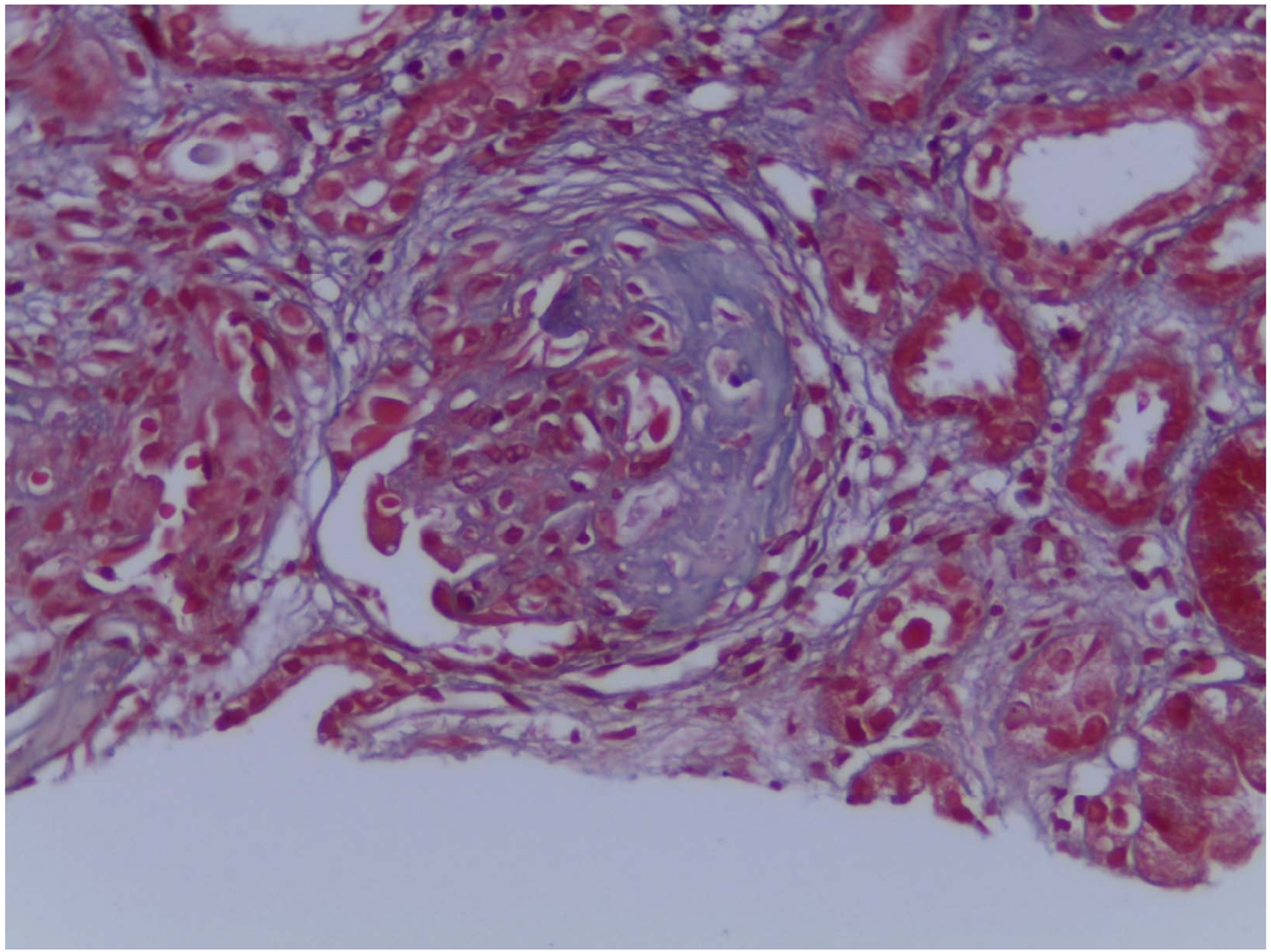

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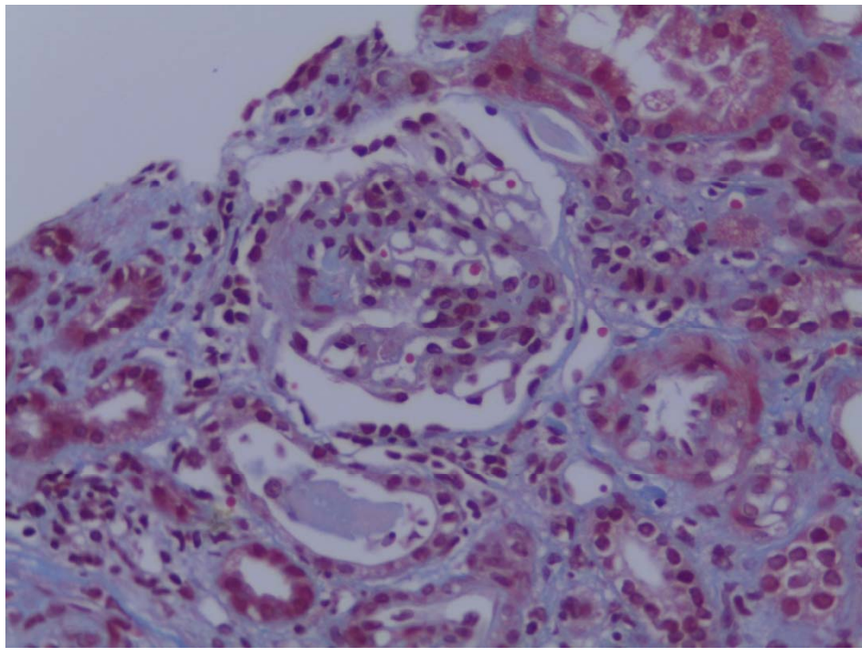

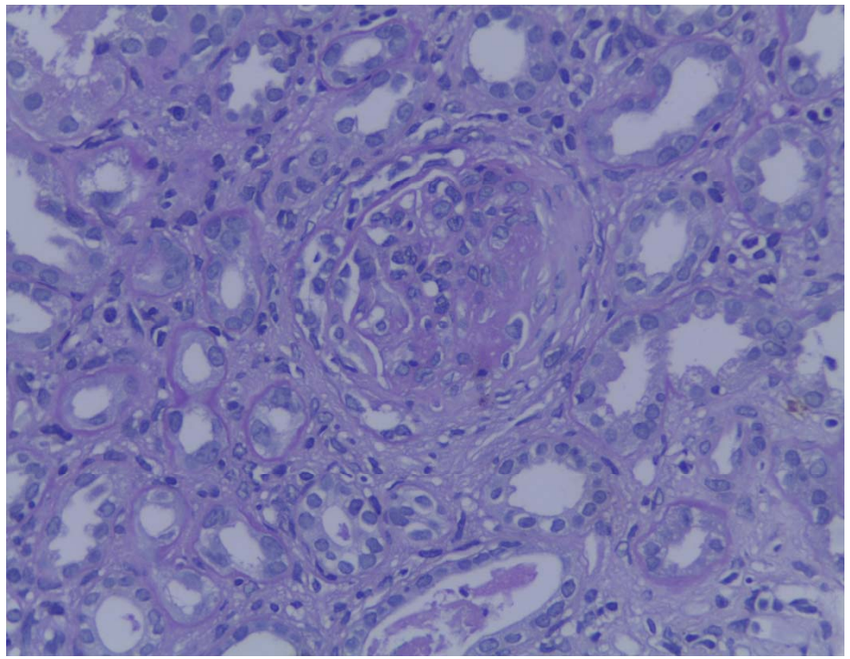

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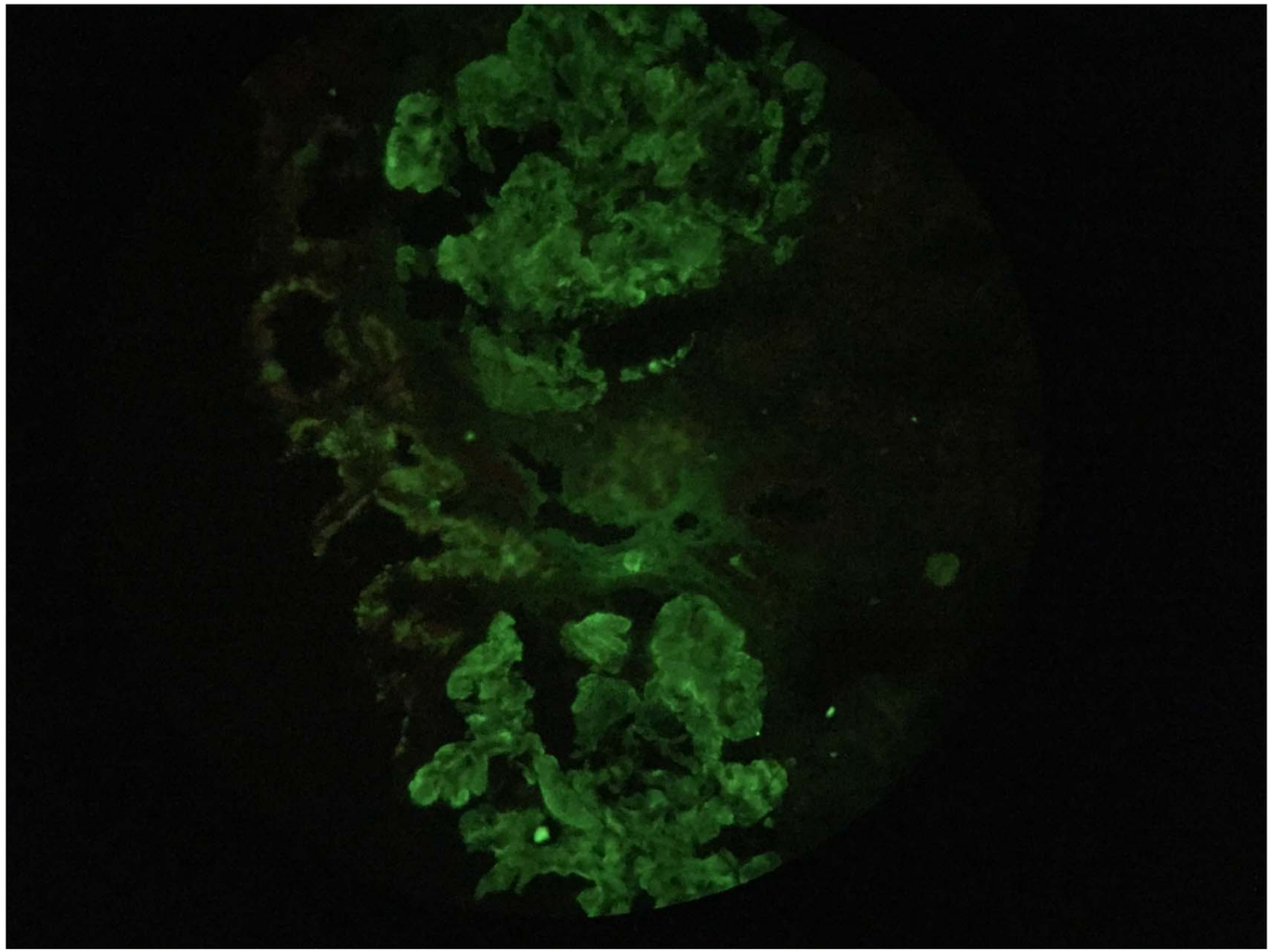

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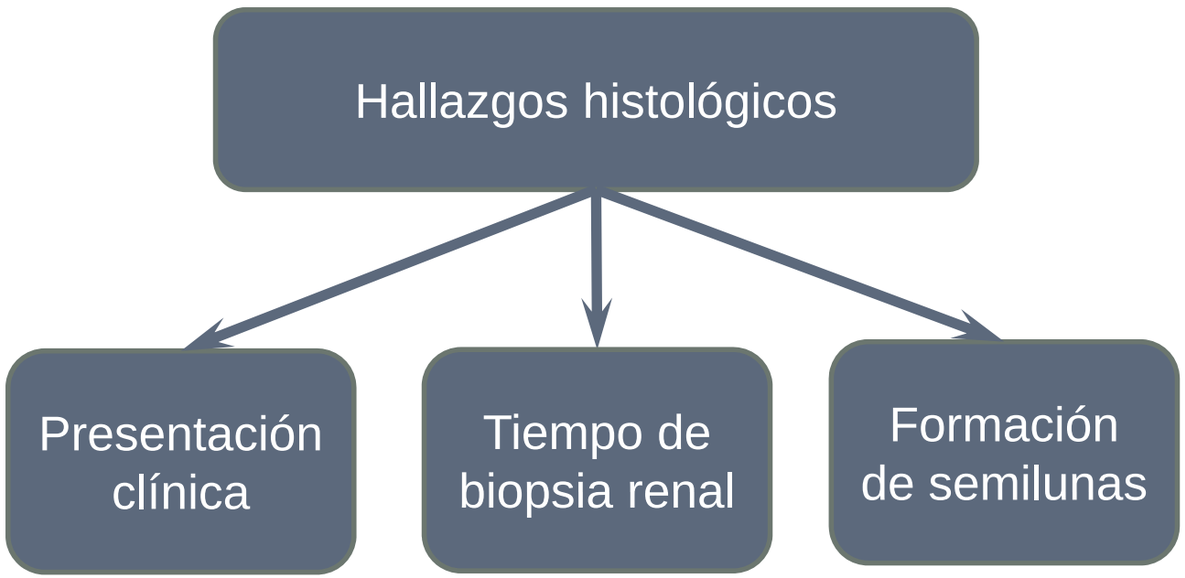

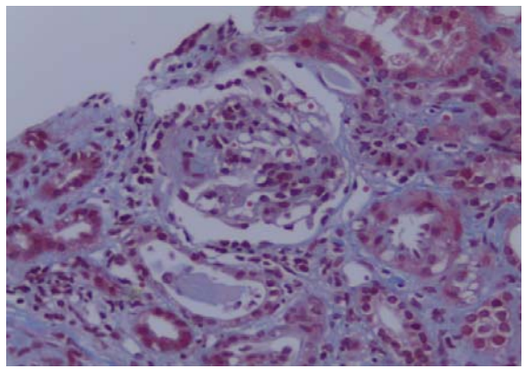

Pediatr Nephrol
DOI 10.1007/s00467-017-3794-1

ORIGINAL ARTICLE

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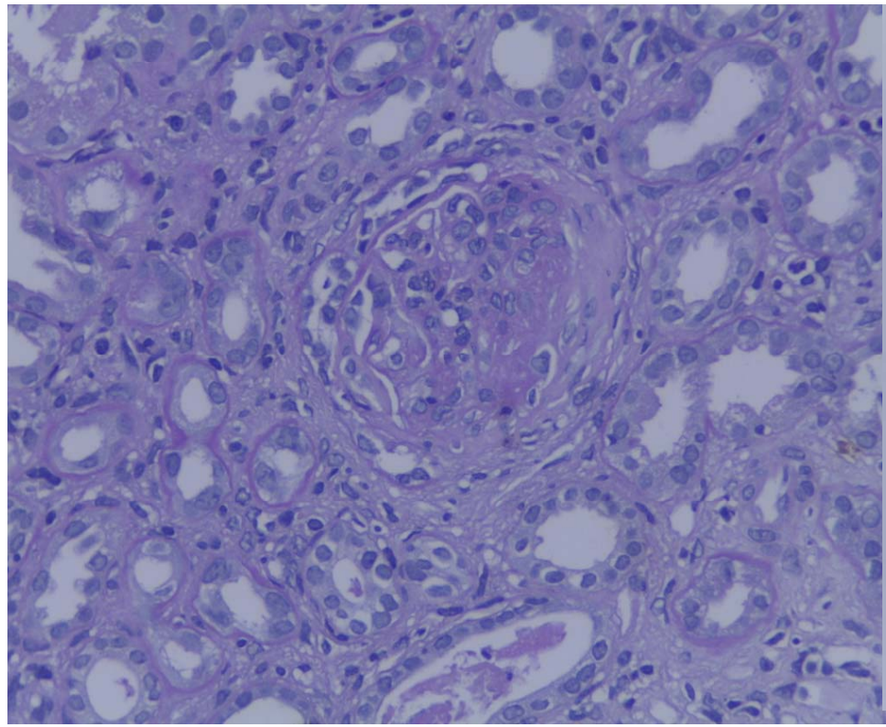
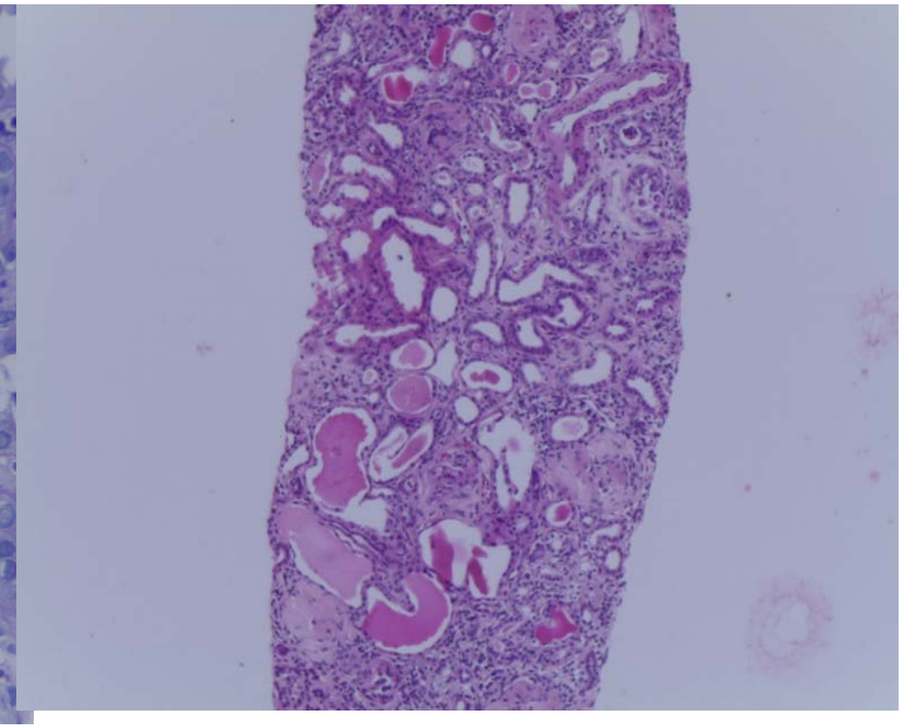
LESIONES CRONICAS EN BIOPSIA RENAL





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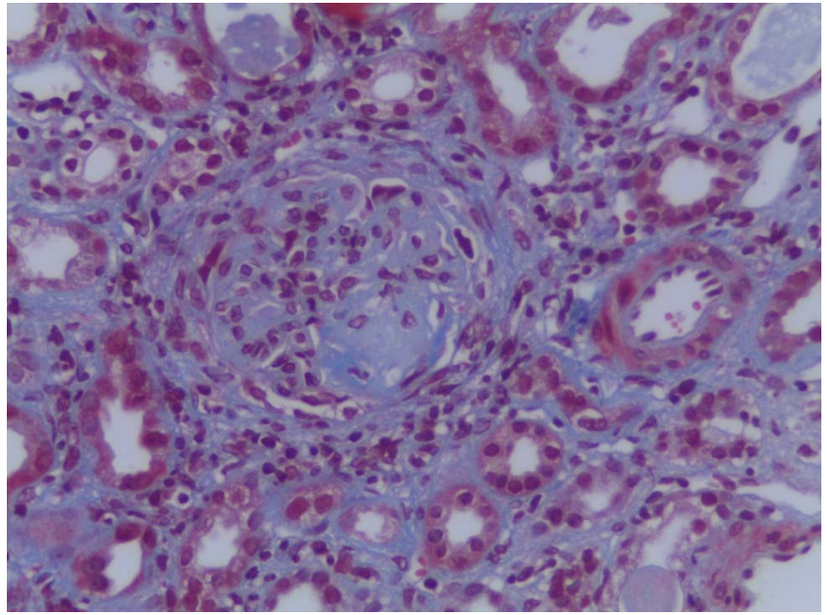
- Biopsia temprana
- Lesiones crónicas pueden desarrollarse en el primer mes del inicio de la enfermedad
- Asociadas a reducción de la FR
- Asociado a proteinura no nefrótica
- Tratamiento temprano reduciría el daño glomerular crónico

BIOPSIA RENAL

Clasificación ISKDC

- No se relaciona completamente con severidad de manifestación clínica y pronóstico a largo plazo.
- No hay diferencia entre lesiones glomerulares crónicas y agudas
- No tiene presente lesiones tubulares e insterticiales



Henoch-Schönlein Purpura Nephritis: Pathophysiology, Treatment, and Future Strategy

Jean-Claude Davin

Clin J Am Soc Nephrol 6: 679–689, 2011. doi: 10.2215/CJN.06710810

Jo
https://doi.org/10.1007/s40620-017-0457-z

ORIGINAL ARTICLE

Value of the Oxford classification of IgA nephropathy in children with Henoch–Schönlein purpura nephritis

Ke Xu[1] · Lili Zhang[2,3] · Jie Ding[1] · Suxia Wang[2] · Baige Su[1] · Huijie Xiao[1] · Fang Wang[1] · Xuhui Zhong[1] · Yanming Li[4]

Received: 16 August 2017 / Accepted: 31 October 2017

BIOPSIA RENAL

- Updated Clasificación Oxford 2017
  (International IgA Nephropathy Network & Renal Pathology Society)
- Hipercelularidad Mesangial (M)
- Proliferación Endocapilar (E)
- Glomeruloesclerosis Segmentaria (S)
- Fibrosis Insterticial / Atrofia Tubular(T)
- Medialunas fibrocelulares / Celulares(C)

GME segmentaria
Atrofia tubular
Fibrosis insterticial

son importante factores pronósticos y no son tenidos en cuenta en vasculitis por IgA

Journal of Nephrology
https://doi.org/10.1007/s40620-017-0457-z

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- Hipercelularidad mesangial → Proteinuria
- Atrofia tubular/ Fibrosis insterticial / semilunas en más del 50% de los glomérulos → Reducción de FG
- Esclerosis glomerular, Atrofia tubular/ Fibrosis insterticial

↓

Importantes factores pronósticos y terapéuticos

TRATAMIENTO

Hematuria microscópica / macroscópica de corta duración /
Proteinuria Menor de 1gr/día

↓

No biopsia

↓

No tratamiento específico

↓

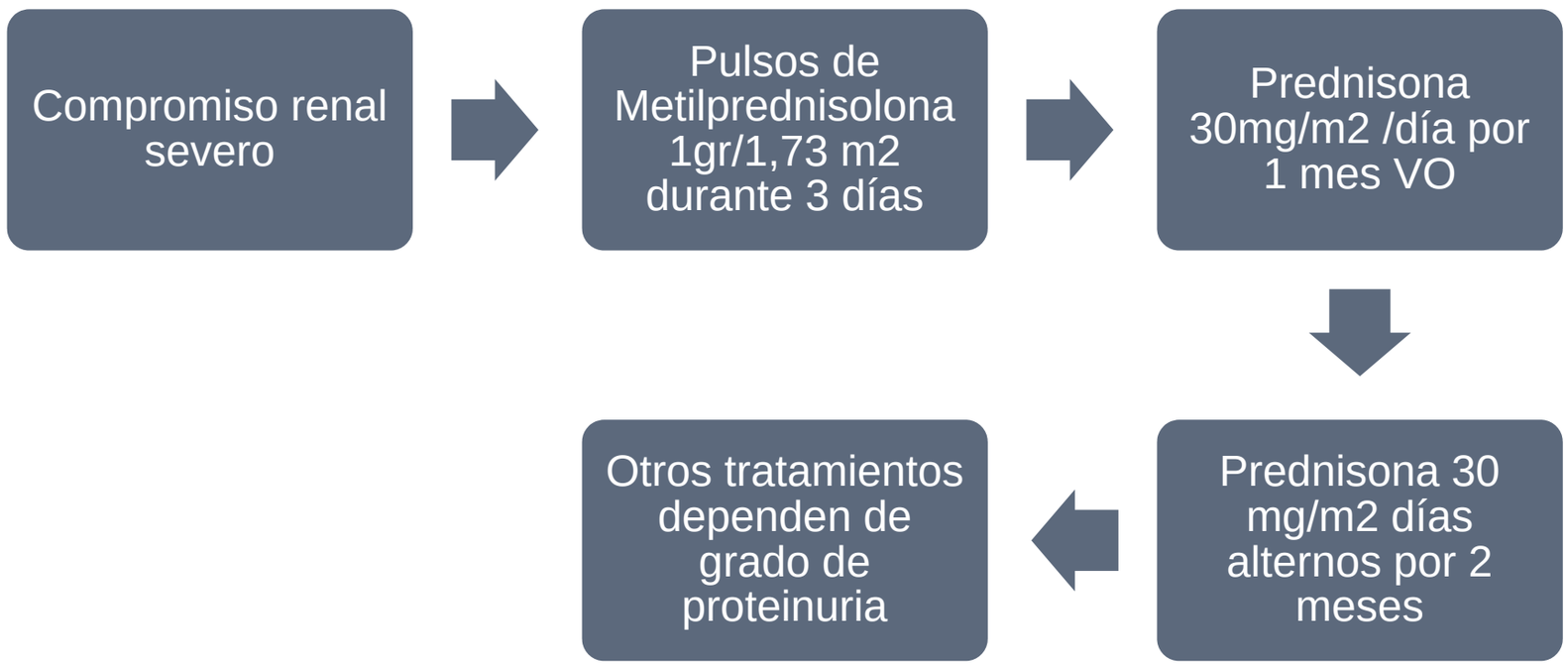
Seguimiento de proteinuria y FR

UpToDate®

IgA vasculitis (Henoch-Schönlein purpura): Renal manifestations

Authors: Patrick Niaudet, MD, Gerald B Appel, MD, Gene G Hunder, MD
Section Editors: Richard J Glassock, MD, MACP, Fernando C Fervenza, MD, PhD
Literature review current through: Mar 2018. | **This topic last updated:** Feb 14, 2018.

TRATAMIENTO



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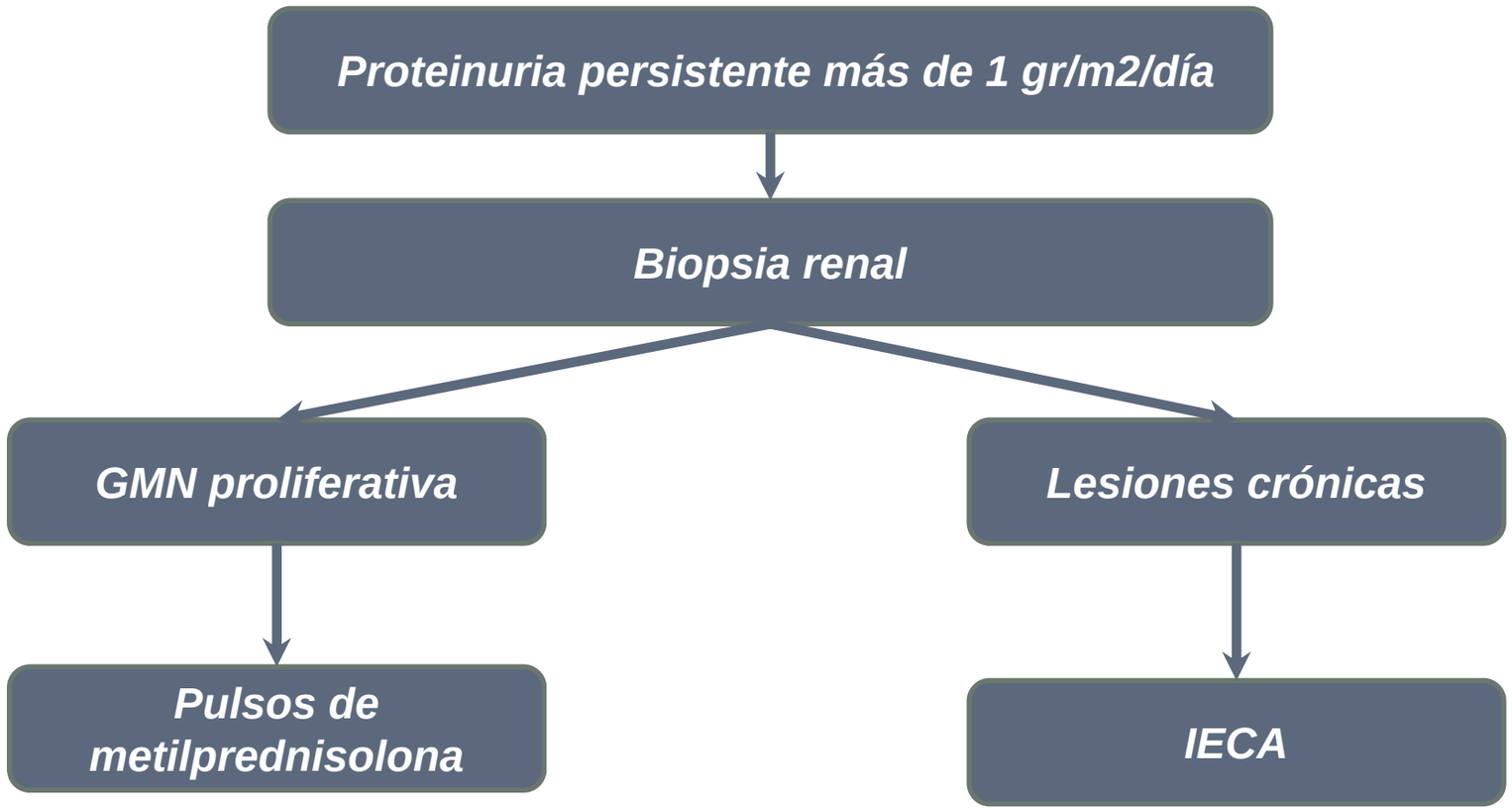

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TRATAMIENTO

- No se recomienda ciclofosfamida, ciclosporina azatioprina, como terapia inicial. Considerar en niños que no responden a glucocorticoides

- No recomendamos tratamiento con glucocorticoides para prevenir la nefritis por vasculitis IgA

UpToDate®

IgA vasculitis (Henoch-Schönlein purpura): Renal manifestations

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CONCLUSIONES

- Las futuras estrategias deberían consistir en mejorar la definición de pacientes con riesgo de enfermedad renal crónica

- La documentación histológica debería considerar todos los factores que pueden aportar información para el tratamiento y pronóstico del niño

MUCHAS GRACIAS