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Underlying Glomerulopathies in a Nationwide Colombian Pediatric Series with Atypical Hemolytic Uremic Syndrome

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Objective

To delineate clinical presentation, histopathological features and outcomes of Colombian pediatric patients with Atypical Hemolytic Uremic Syndrome (aHUS).

Methods

This multicenter cohort enrolled 27 Colombian children with aHUS (2010-2019). Patients grouped by age at onset. Clinical features compared using ANOVA/Fisher exact tests. Renal biopsy performed on six patients initially suspected of other renal diseases.

Results

Mostly male patients (70%) had aHUS onset before age 4 (60%), triggered mainly by gastroenteritis (52%). Pulmonary involvement (67%) was more frequent in the 1-7 age group (p=0.01) (Figure 2) Biopsies showed 3 MPGN type I, 1 MPGN type III, 1 C3GN, and 1 RPGN (Table 1) (Figure 1). Genetic screening in 5 patients identified 2xCFHR5, 2xMCP, and 1xADAMTS-13/THBD mutations. 15 relapses occurred, with 8 (72%) in 1-7 age group. Renal outcomes were similar across age groups.

Conclusion

Extrarenal involvement was frequent at presentation, particularly with pulmonary manifestations. The histopathological features support the alternative pathway hyperactivation mechanism in MPGN, C3GN and aHUS

Table 1. Clinical data of six pediatric aHUS patients with biopsy-proven glomerulopathy

ID	Age (years)	Sex	Glomerulopathy	IF (C3)	IF (IgG)	24-h PU (g)	Serum C3 (>80 mg/dl)	Serum C4 (>15 mg/dl)	Focused Therapy	Outcome
2	15	Male	MPGN I	+++	+	0,4	37	17	PE, steroids	ESKD
3	12	Male	MPGN I	+++	+	0.8	45	32	PE, steroids	ESKD
7	0,3	Male	MPGN I	+++	+	2.2	46	16	Eculizumab	Slight PU
11	3	Male	MPGN III	++	-	3.4	56	19	Eculizumab	ESKD, Died
19	0,2	Female	RPGN	-	-	0.4	132	30	PE, steroids	ESKD, Died
20	1	Male	C3GN	+++	-	0.9	28	24	Eculizumab	Slight PU

CKD, chronic kidney disease; ESKD, end-stage kidney disease; FSGS, focal and segmental glomerulosclerosis; C3GN, C3 glomerulonephritis; IF, immunofluorescence staining; MPGN, membranoproliferative glomerulonephritis; RPGN: Rapidly progressive glomerulonephritis; PE, plasma exchange; PU, proteinuria.

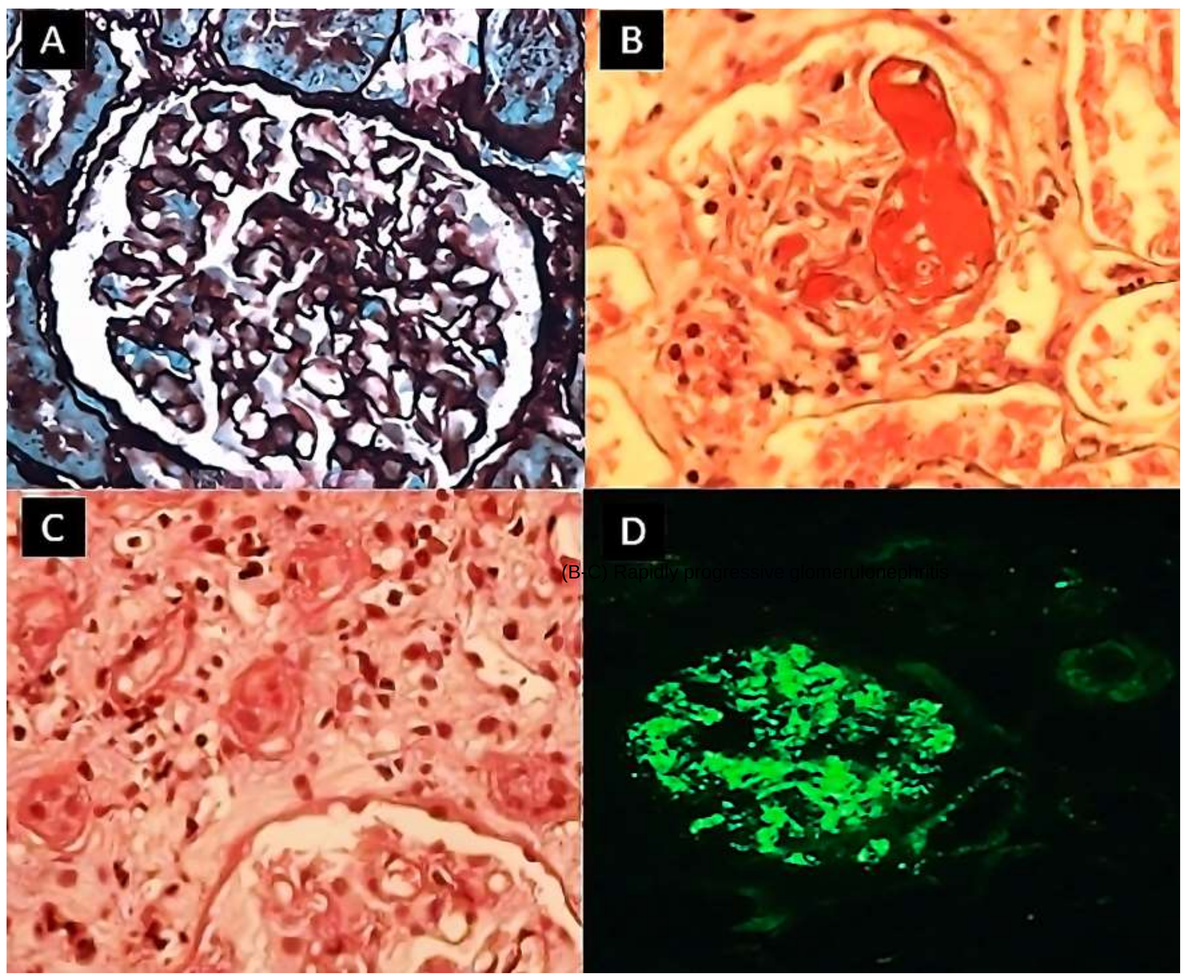

Figure 1: Light microscopy findings in children who developed glomerulopathy associated with aHUS

(A) Membranoproliferative glomerulonephritis

(B-C) Rapidly Progressive glomerulonephritis

(B) Thrombotic Microangiopathy
(C) Endothelial Proliferation and Fibrinoid Microthrombi

(D) C3 Glomerulopathy

(Anti-C3c antibody × 400)
Diffuse granular deposition

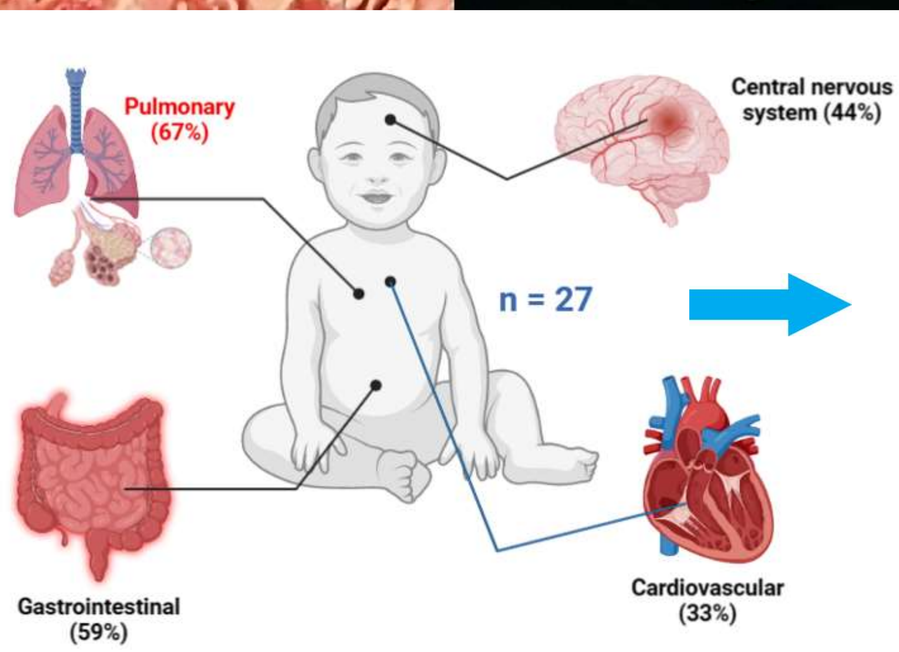

Figure 2: Extrarenal involment in Colombian Pediatric patients with Atypical Hemolytic Uremic Syndrome

Pulmonary manifestations were the most frequent extrarenal involment

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