Abstract 364 Table 1 Showing comparison of baseline parameters in both treatment groups.

Parameter	Deflazacort Group (n=31)		Prednisolone Group (n=29)	
Age in years (mean±SD)	27.3±9.3		28.3±7.6	
Sex	All female		All female	
Duration of Steroid Intake before study entry (mean in days±SD)	212.9±280		124.8±106.1	
Steroid dose in before study entry (mean in mg/day±SD)	14.3±20		11.3±15.9	
Indications for treatment- n(%)	LN Class II	1 (3.33%)	LN Class II	0(0)
	LN Class III	1(3.335)	LN Class III	3(10.3%)
	LN Class IV	12(38.7%)	LN Class IV	11(37.9%)
	LN Class V	3(9.7%)	LN Class V	4(13.7%)
	CNS Lupus	4 (12.9%)	CNS Lupus	2(6.9%)
	Vasculitis	4(12.9%)	Vasculitis	4(13.7%)
	Cytopenias	3 (9.7%)	Cytopenias	3(10.3%)
	Hemolytic Anemia	1 (3.33%)	Hemolytic Anemia	1(3.5%)
	Myositis	1(3.33%)	Myositis	0(0)
	ILD	1(3.33%)	ILD	0(0)
	Autoimmune Hepatitis	0(0)	Autoimmune Hepatitis	1(3.5%)

Conclusions DFZ and PDN used in comparable manner in SLE had similar efficacy with significantly lesser weight gain, lesser cushingoid features (including lesser glycaemic elevation) seen in DFZ group.

365 PROFILE OF HENOCH SCHONLEIN PURPURA (HSP) NEPHRITIS: 23 YEARS EXPERIENCE AT A TERTIARY CARE CENTRE IN NORTH INDIA

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10.1136/lupus-2017-000215.365

Background and aims Henoch Schonlein Purpura (HSP) is one of the most common vasculitides of childhood. Glomerulonephritis is seen in approximately 30%–50% of the patients and is the principal cause of morbidity and mortality in HSP patients.

Methods 314 children were diagnosed with HSP from 1993–2015 based on EULAR/PRINTO/PRES criteria. A retrospective case review of all patients with HSP Nephritis (HSPN) was done. HSPN was defined based on urine erythrocyte >5/HPF and proteinuria. Patients were divided into four clinical types (Table 1). The severity of renal pathological findings was determined based on the classification of International Study of Kidney Disease (ISKDC), from grades I – VI.

Results Renal involvement was seen in 64 patients after a mean duration of 32.3 days from the onset of symptoms of HSP. Details of patients with HSPN is summarised in table 2, 3 and figure 1. Three fourth of the patients had histological grade II or IIIa (figure 2). 75% of patients with grade $\geq$ IV had gross hematuria at presentation. Treatment details are shown in figure 3. Patients were followed up for a mean period of 42.9 months during which 13 were lost to follow up and 1 expired. Nephritis resolved in 48 patients (75%). 13 patients developed renal relapse manifesting as albuminuria with microscopic hematuria in 77% patients followed by isolated albuminuria (23%).

Conclusions Renal involvement was noted in 20.4% of children with HSP. Massive proteinuria was the most common clinical feature. Grade II and IIIa were the most common renal pathological grades.

366 SPLICING FACTOR PROLINE/GLUTAMINE-RICH (SFPQ) IS A NOVEL AUTOANTIGEN OF ANTI-MDA5 ANTIBODY-POSITIVE DERMATOMYOSITIS/CLINICALLY AMYOPATHIC DERMATOMYOSITIS

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10.1136/lupus-2017-000215.366

Background and aims Anti-MDA5-positive dermatomyositis (DM) and clinically amyopathic DM (CADM) often develop

Abstract 364 Table 2 Showing Δ Changes for Outcome parameters concerning Primary Objectives (in terms absolute and percentege change) from baseline to the follow up visits.

Parameter	Deflazacort Group (mean±SD)		Prednisolone Group (mean±SD)		p-value (absolute,% change)
	Absolute change	% Change	Absolute Change	% Change	
Δ Weight in kilograms 1st-2nd visit	1.9±1.7(n= 31)	3.9±3.6	3.8±2.4 (n=29)	8.0±5.4	0.001,0.001
Δ Weight in kilograms 1st-3rd visit	2.5±2.8 (n=29)	5.1±5.9	5.9±3.7 (n=29)	12.4±7.6	0.001,0.001
Δ Hirsuitism Index 1st- 2nd visit	0.9±0.8 (n=31)	9.2±9.0	2.00±1.1 (n=29)	21.8±13.0	0.001,0.001
Δ Hirsuitism Index 1st- 3rd visit	1.4±1.2(n=29)	14.7±13.1	3.0±1.2 (n=29)	33.3±13.9	0.001,0.001
Δ CSI 1st- 2nd visit	1.1±1.0 (n=31)	82.3±79.8	1.7±1.3 (n=29)	138.5±131.9	0.04,0.129
Δ CSI 1st- 3rd visit	1.4±0.9 (n=29)	72.3±81.6	2.4±1.3 (n=29)	171.2±141.2	0.002,0.01
Δ FBS in mg/dl 1st- 3rd visit	-5.6±17.9 (n=29)	-4.2±18.2	5.5±18.4(n=29)	7.6±20.8	0.02,0.01
Δ PPBS mg/dl 1st- 3rd visit	-2.3±27.8(n=29)	1.4±20.2	14.7±38.2(n=29)	15.8±34.40	0.05,0.15
Δ HbA1C1st- 3rd visit (% DCCT units)	-0.03±0.4(n=29)	-3.5±20.0	0.2±0.7(n=29)	5.1±13.8	0.11,0.06

rapidly progressive interstitial lung disease, but their pathogenesis remain unclear. We observed that sera from anti-MDA5-positive DM/CADM patients immunoprecipitated a common 110 kDa polypeptide. We investigated the autoantigen and its clinical significance.

Methods Autoantibodies were screened in 340 patients with various connective tissue diseases (CTDs) and 20 healthy controls (HCs) by immunoprecipitation with [^{35}S]methionine-labelled HeLa cells. Immunoabsorbent column chromatography was used to purify the reactive autoantigen which was subsequently analysed by peptide mass fingerprinting.

Results Anti-110 kDa was detected in sera from 25 DM/CADM patients with anti-MDA5 but not in those from other CTDs or HCs. All anti-MDA5-positive DM/CADM patients who showed the recurrence symptoms had anti-110kDa. Interestingly, all who required plasma exchange were not positive for anti-110kDa at the initial plasma exchange. The corresponding autoantigen was identified as splicing factor proline/glutamine-rich protein (SPFQ). In some cases, anti-SFPQ was detected at the diagnosis (early-group), but in other cases, it appeared during the disease course (delayed-group). The diagnosis time of DM/CADM had seasonal patterns according to the temporal appearance of anti-SFPQ antibody. 67% (8/12) of patients were diagnosed between August and October in the early-group, whereas 58% (7/13) of patients were between January and March in the delayed-group.

Conclusions SFPQ is a novel autoantigen of anti-MDA5-positive DM/CADM. The diagnosis timing of DM/CADM with anti-MDA5 have seasonal patterns according to the appearance timing of anti-SFPQ. These findings may provide new insights into the pathogenesis of DM/CADM.