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# Oral presentations 1

## Electron microscopy — Neuropathology — Miscellaneous

### OP1.1

### Immunohistochemical and electron-microscopic study in Henoch-Schoenlein nephritis

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**Background** Henoch-Schoenlein purpura is a form of systemic vasculitis with deposits of IgA, involving small vessels in skin, gut and glomeruli. Henoch-Schoenlein nephritis (HSN) is the most common secondary childhood nephropathy and leads to ESRD in up to 20% of pediatric patients after 20 years of follow-up.
**Methods** 44 cases of HSN: 32 children and 12 adults. EM was performed in 6 cases and IH in all. We evaluated mesangial proliferation (Ki-67, PCNA), mesangial expression of α-SMA and podocytes expression of p27.
**Results** LM study revealed the following grades: II (18), III (15), IV (3) and VI (8) (differences in prognosis between grades were not found). The initial symptoms and outcome were less severe in patients with grade II or III. Presence of glomerular sclerosis was important prognostic marker. EM study was was useful in recognition of early glomerular sclerosis. No significant correlations were found between mesangial cells Ki-67 and PCNA positivity and clinical presentation. Important finding was progressive decrease in p27 positive podocytes with more severe HSN grades. Significant correlations regarding the area of mesangial expression of α-SMA were observed. That expression was stronger in patients, who had foci of infection (independently of HSN grade).
**Conclusion(s)** The area of α-SMA expression have proved to be a useful marker of mesangial cells activation. The decreased expression of p27 in podocytes of HSN patients suggests their role in disease process. Our study did not demonstrated the prognostic usefulness of proliferation markers (Ki-67, PCNA).

### OP1.2

### Remodeling of adipose tissue from lipodystrophic patients with LMNA mutations

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**Background** A-type lamins, encoded by the LMNA gene, are ubiquitous nuclear intermediate filament proteins that are required for the structural and functional integrity of the nucleus. Inherited laminopathies, due to LMNA mutations, represent a wide spectrum of diseases, including lipodystrophies, with peripheral subcutaneous fat loss, increase visceral fat and metabolic alterations; among them, the Dunningan-type familial partial lipodystrophy (FPLD2; OMIM 151660). The morphological alterations of adipose tissue in laminopathies have not been reported so far. In the present work, we have studied the pathological and ultrastructural alterations of adipose tissue from LMNA-mutated patients, together with the expression of specific adipocytes markers and mitochondrial proteins.
**Methods** Cervical fat was obtained from three patients with LMNA mutation, from one patient with mtDNA tRNA lys A8344G mutation, from three treated lipodystrophic HIV-infected men and three controls. Light microscopy, immunohistochemical and ultrastructural studies were performed on the fat samples and compared with mRNA assay, performed with the light cycler software (Roche diagnostics) and Western blot analysis.
**Results** Adipose tissue from LMNA-mutated patients shows a heterogeneous structure with decreased adipose size, altered extra-cellular matrix : thick fibrils invade the intercellular area without macrophages or inflammation, increased fibrosis, mitochondria disturbances, accumulation of prelamin A and altered expression of adipogenic transcription factors.
**Conclusion(s)** This study show here severe remodeling of adipose tissue from patients with FPLD and metabolic laminopathies. The accumulation of prelamin A could impair SREBP1 function and adipocyte differentiation but also induce oxidative stress leading to fibrosis.