Disclosure of Interest
None declared

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Lupus nephritis in childhood: the analysis of a cohort in the transitional age
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Introduction: Lupus nephritis (LN) is the onset of systemic lupus erythematosus (SLE) in 50-80% of children; otherwise, LN appears, in most patients, in the first two years of illness. Some patients have nephritis as the only manifestation of SLE. Histological diagnosis confirms renal involvement, defines the damage extent, guides treatment, based on the six classes of nephritis.
Objectives: The aim of our work is to compare the effectiveness of the different therapeutic schemes for LN in a single center cohort, to ensure the appropriate time for transition from the paediatric to the adult Rheumatology care.
Methods: A retrospective analysis of SLE patients followed in the period 2000-2023, selected 9 patients (8F;1M: age: 9-16years); mean age at the LN onset: 12 years.
Results: LN was the first sign of SLE in 5/9 (56%), after 0.5-2 years in 4 (44%). At the onset, 100% presented proteinuria >500 mg/24h, in 2 with microhaematuria. 6/9 (66%) had a nephrotic syndrome, 4 of whom needed haemodialysis treatment for acute renal failure. One patient presented antiphospholipid antibody syndrome with thrombotic manifestations. Four (44%) had hypertension. Positive autoantibodies (ANA, ENA, anti-dsDNA, anti-nucleosome) were detected in 8/9 (88%), hypocomplementemia in 4/9 (44%). The diagnosis of LN was confirmed by histology: 6 (67%) had class IV, 3 (33%) class V nephritis. All patients were treated with intravenous methylprednisolone boluses, followed by oral prednisone, with a slow dose tapering at the remission. Induction therapy, in patients with diffuse proliferative glomerulonephritis, was done with cyclophosphamide, in combination with steroid (1-2 mg/kg/day) in 5/6 (83%), with mycophenolate in 1. 8/9 (88%) received maintenance treatment with mycophenolate; 1 patient with cyclosporine, obtaining in most clinical improvement and normalization of proteinuria. One patient, non-responder to therapy, needed haemodialysis treatment and inclusion in the kidney transplant list. All patients with membranous glomerulonephritis received mycophenolate in induction and in maintenance, except 1, treated in induction with prednisone and azathioprine. The analysis of the follow-up data showed lower frequency of relapses with mycophenolate. The transitional care was started only for patients in remission, to a Rheumatology center with a multispecialistic team.
Conclusion: Our patients received a different therapeutic approach, depending on the year of the diagnosis. Our case series confirms the importance of LN histological diagnosis, to personalize therapy and improve quality of life in the perspective of transition (1). It is desirable that the patient faces the transition into a phase of remission of the disease, to a Rheumatology center with a multispecialistic team, to abate relapses risk and to improve long-term outcome.

Patient Consent
Yes, I received consent

Disclosure of Interest
None declared

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Effect of immunomodulatory therapies on antiphospholipid antibodies titers in children with antiphospholipid syndrome
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Introduction: Pediatric Antiphospolipid Syndrome (APS) is an autoimmune disease characterized by thrombotic events (TE) associated with 2 consecutive positive determinations of antiphospholipid antibodies (aPL) [1]. High levels of aPL are associated with increased thrombotic risk. The use of immunomodulatory therapies with the aim of reducing aPL titers may represent a treatment strategy. Therefore, monitoring of aPL levels may represent a strategy to evaluate both disease activity and response to therapies.
Objectives: To investigate the trend over time of aPL titers in children with APS, comparing patients under immunomodulatory therapies and those without them.
Methods: A descriptive, observational, cross-sectional study was carried out in children with APS. aPLs were tested from diagnosis every 3-4 months for 2 years. Interferon Gene Signature (IGS) was also assessed. Laboratory, clinical and demographic data was retrieved and analyzed.
Results: Sixteen children with a diagnosis of APS were included. The median age at disease onset was 11.5 years (range: 6 months – 17 years); 63% were girls; 88% Caucasians. Thirteen patients (81%) had a diagnosis of primary APS and 3 (19%) secondary APS.
Regarding clinical manifestations, 11 children developed at least one TE (7 arterial and 5 venous). Cerebral territory was the most frequently involved (5), followed by 3 deep vein thrombosis , 1 pulmonary thromboembolism and 2 arterial renal thrombosis. 13 and 8 patients received an antiaggregant and/or anticoagulant therapy, respectively. Twelve (75%) children developed at least one non-criterion manifestation, 44% of which were cardiac (Libman-Sacks endocarditis or valvular heart disease), 44% neurological (chorea or white matter lesions), 38% hematological (thrombocytopenia, hemolytic anemia or Evans syndrome) and 4% renal (thrombotic microangiopathy).
The rates of aPLs recorded were: 25% single positivity, 42% double and 44% triple; showing the highest rates IgG aβ2GP (88%) and IgG aCL (81%). Lower rates were identified for LA (44%), IgM aCL (19%) and IgM aB2GP (13%). Four (25%) were ANA positive (3 secondary APS).
Regarding the therapy, 13 children (81%) received at least one immunomodulatory drug (13 mycophenolate; 4 rituximab) and 3 (19%) did not receive any treatment.
During the 2-year-follow-up, 11 patients (69%) showed a reduction of aPL titers, becoming 9 (56%) negative (Fig 1). Of those who were negative, 8 received an immunomodulatory therapy. Five patients (31%) showed stable titers during follow-up. Of them, 2 were not treated and 2 were under mycophenolate but with poor compliance (a reduction of titers with therapy resumption was observed).
IGS at diasease onset was also evaluated in 11 patients, resulting positive in 9 (82%).
Conclusion: Our data suggest the possible effect of immunomodulatory therapies in reducing antibody titers in APS. Therefore, it may represent a strategy to control the disease activity leading to a better prognosis. Further studies are needed to confirm and expand our data.

Patient Consent
Yes, I received consent