O44. AN INTEGRATIVE ANALYTICAL APPROACH TO SUBPHENOTYPING OF JUVENILE DERMATOMYOSITIS
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Background: JDM is a severe paediatric autoimmune condition associated with muscle weakness and skin rashes, affecting approximately 1 in 20 000 children. It encompasses a heterogeneous spectrum of symptoms, and can involve differing degrees of muscle weakness, treatment-resistant rash, calcinosis, ulceration and involvement of the lung, gut or brain. Existing treatment consists of long-term management involving steroids and immunosuppression; while some patients are responsive and able to come off treatment by 2 years, others fail to respond. In order to improve management of JDM, there is an imperative to define clinical subphenotypes that are identifiable by unique biomarkers, and to investigate the biological mechanisms underpinning these subtypes. To that end, associations have been identified between certain clinical features and expression of the anti-MDA5, anti-NXP2 and anti-TIF1-γ autoantibodies.

Methods: Muscle biopsies (n = 101) from the UK-wide JDM Cohort and Biomarker Study were scored using the validated score tool. Biopsy scores are being integrated with clinical features at the time of biopsy and at subsequent time-points. Principal component analysis (PCA) has been performed as a clustering technique in order to define JDM sub-phenotypes.

Results: Preliminary PCA has successfully identified two distinct clusters that correlate with the autoantibodies anti-MDA5 and anti-Mi2, and reflect mild and severe histological changes, respectively. Separation between these clusters was predominantly on the basis of biopsy features in the Inflammatory and Muscle Fibre Domains in the score tool.

Conclusion: These analyses represent the first step towards the identification of JDM sub-phenotypes that correlate with potential biomarkers, and may enable definition of biological mechanisms behind such subtypes and a more targeted therapeutic approach for JDM.

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