The new classification criteria for paediatric vasculitis

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Until recently paediatrician have relied on classification criteria developed by considering patient groups which did not include children. Although children/adolescents and adults with vasculitis share many signs and symptoms of disease, they differ in the relative frequency of some clinical manifestations and concomitant diseases. Therefore, it cannot be assumed a priori that the classification criteria developed for adults are suitable for children and adolescents.

In 2005 the vasculitis working group of the Paediatric Rheumatology European Society (PRES) proposed preliminary classification criteria, for some of the most frequent childhood vasculitides [10] namely Henoch-Schönlein purpura (HSP), childhood polyarteritis nodosa (c-PAN), c-Wegener granulomatosis (c-WG), c-Takayasu arteritis (c-TA). Subsequently with support from European League Against Rheumatism (EULAR), the Paediatric Rheumatology International Trials Organization (PRINTO) and PRES, PRINTO/PRES established a formal statistical validation process with a large scale data collection that culminated in the final 2008 Ankara Consensus Conference.

With this first review we describe the general methodology and overall clinical, laboratory, and radiographic characteristics, and the specific details on the final classification criteria for each of the 4 vasculitides analysed (HSP, c-TA, c-PAN and c-WG).

The validation project was divided into 3 main steps. Step 1: retrospective/prospective web-data collection of HSP, c-PAN, c-WG and c-TA, with age at diagnosis ≤ 18 years. Step 2: blinded classification by consensus panel of a subgroup of 280 cases (128 difficult cases and 152 randomly selected) enabling expert diagnostic verification. Step 3: Ankara 2008 consensus conference and statistical evaluation (sensitivity, specificity, AUC, k-agreement) using as gold standard the final consensus classification or original treating physician diagnosis.

A patient is classified as HSP in the presence of purpura or petechiae (mandatory) with lower limb predominance plus 1/4 criteria: 1) abdominal pain, 2) histopathology (IgA), 3) arthritis or arthralgia, 4) renal involvement. Classification of c-PAN requires a systemic inflammatory disease with evidence of necrotising vasculitis OR angiographic abnormalities of medium/small sized arteries (mandatory criterion) plus 1/5 criteria: 1) skin involvement, 2) myalgia/muscle tenderness, 3) hypertension, 4) peripheral neuropathy, 5) renal involvement. Classification of c-WG requires 3/6 criteria: 1) histopathologic evidence of granulomatous inflammation; 2) upper airway involvement; 3) laryngo-tracheo-bronchial involvement; 4) pulmonary involvement (X ray/CT); 5) ANCA positivity; 6) renal involvement. Classification of c-TA requires typical angiographic abnormalities of the aorta or its main branches and pulmonary arteries (mandatory criterion) plus 1/5 criteria: 1) pulse deficit or claudication, 2) blood pressure discrepancy in any limb, 3) bruits, 4) hypertension, 5) elevated acute phase reactant.

This methodological approach allow EULAR/PRINTO/PRES to propose validated classification criteria for HSP, c-PAN, c-WG and c-TA.