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### Table III. Definitions of ANCA-patterns

#### **P-ANCA**

Consensus:<sup>1</sup> perinuclear fluorescence, with or without nuclear extension

Alternatives:<sup>2</sup>

- Perinuclear fluorescence with nuclear extension on ethanol-fixed neutrophils (i.e. excluding X-ANCA)
- Perinuclear fluorescence, with or without nuclear extension on ethanol-fixed neutrophils and MPO-ANCA in antigen-specific immunoassay
- Perinuclear fluorescence, with or without nuclear extension on ethanol-fixed neutrophils and classic granular cytoplasmic fluorescence with central or interlobular accentuation on formalin-fixed neutrophils
- Perinuclear fluorescence, with or without nuclear extension on ethanol-fixed neutrophils, classic granular cytoplasmic fluorescence with central or interlobular accentuation on formalin-fixed neutrophils, and MPO-ANCA in antigen-specific immunoassays

#### **Atypical ANCA**

Consensus:<sup>1</sup> all neutrophil-specific IIF reactivity other than C-ANCA, atypical C-ANCA, or P-ANCA; most commonly a combination of cytoplasmic and perinuclear fluorescence

Alternatives:<sup>2</sup>

- Perinuclear fluorescence without nuclear extension on ethanol-fixed neutrophils (also referred to as X-ANCA)
- Perinuclear fluorescence, with or without nuclear extension on ethanol-fixed neutrophils and negative on formalin-fixed neutrophils
- Any neutrophil-specific IIF reactivity on ethanol-fixed neutrophils and negative on formalin-fixed neutrophils
- Any neutrophil-specific IIF reactivity that is not confirmed as MPO- and/or PR3-ANCA in antigen-specific immunoassays
- Diffuse homogeneous cytoplasmic fluorescence without typical interlobular accentuation on ethanol-fixed neutrophils (i.e. consensus definition of atypical C-ANCA)
- Antinuclear antibody reactivity

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<sup>1</sup>Consensus patterns are defined on IIF reactivity on ethanol-fixed neutrophils

<sup>2</sup>Only most prevalent alternatives are listed. In some countries also reactivity on methanol-fixed neutrophils is included in the definition. In general, antinuclear antibody interference is to be excluded.