

## 162. CLUSTER ANALYSIS TO EXPLORE CLINICAL SUBCLASSIFICATION OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG-STRAUSS)

Emma Rubenstein<sup>1</sup>, Carla Maldini<sup>1</sup>, Augusto Vaglio<sup>2</sup>, Paolo Bottero<sup>3</sup>, Phillip Bremer<sup>4</sup>, Giacomo Emmi<sup>5</sup>, Paolo Fraticelli<sup>6</sup>, Andrea Gioffredi<sup>2</sup>, Gina Gregorini<sup>7</sup>, Julian Grosskreutz<sup>8</sup>, Giuseppe Guida<sup>9</sup>, Bernhard Hellmich<sup>10</sup>, Claus Kroegele<sup>8</sup>, Federica Maritati<sup>11</sup>, Alberto Pesci<sup>12</sup>, Matthieu Resche-Rigon<sup>1</sup>, David Saadoun<sup>13</sup>, Renato Sinico<sup>11</sup>, Barbara Sokolowska<sup>14</sup>, Wojciech Szczeklik<sup>14</sup>, Jochen Zwerina<sup>15</sup>, Frank Moosig<sup>16</sup>, Thomas Neumann<sup>17</sup> and Alfred Mahr<sup>1</sup>

<sup>1</sup>Saint Louis Hospital Paris, France, <sup>2</sup>University Hospital of Parma, Italy, <sup>3</sup>Ospedale di Magenta Milano, Italy, <sup>4</sup>Clinical Center Bad Bramstedt Bad Bramstedt, Germany, <sup>5</sup>University of Firenze, Florence, Italy, <sup>6</sup>Ospedali Riuniti Ancona, Italy, <sup>7</sup>Spedali Civili Brescia Brescia, Italy, <sup>8</sup>University Hospital of Jena Jena, Germany, <sup>9</sup>Birago di Vische Hospital Torino, Italy, <sup>10</sup>Vasculitis Center Tübingen-Kirchheim Kirchheim unter Teck, Germany, <sup>11</sup>University of Parma Parma, Italy, <sup>12</sup>University of Milano Bicocca Milan, Italy, <sup>13</sup>Pitié-Salpêtrière University Hospital Paris, France, <sup>14</sup>Jagiellonian University Medical College Krakow, Poland, <sup>15</sup>Hanusch Hospital Vienna, Austria, <sup>16</sup>Rheumazentrum Schleswig-Holstein Mitte Neumünster, Germany, <sup>17</sup>Kantonsspital St. Gallen St Gallen, Switzerland

**Background:** Previous studies of eosinophilic granulomatosis with polyangiitis (Churg–Strauss, EGPA) suggested that EGPA may involve clinically distinct phenotypes. In particular, clinical manifestations might be determined by the presence or absence of antineutrophil cytoplasmic antibodies (ANCA) and reflect more prominent underlying vasculitic than eosinophilic disease processes. This study explored whether EGPA involves several subcategories by using hierarchical cluster analysis.

**Methods:** This study used clinical data for patients diagnosed with EGPA in 16 centers in Austria, France, Germany, Italy and Poland collected with a standardized case report form. The analyses pertained to patients reported to have asthma and who fulfilled the Lanham and American College of Rheumatology criteria, with ANCA positivity (by immunofluorescence or ELISA test) as an additional item. We used a multiple correspondence analysis followed by hierarchical cluster analysis with the Ward method. The cluster model included 10 clinical variables assessed at diagnosis (arthromuscular, mucocutaneous, ophthalmological, ENT, cardiovascular, pulmonary, gastrointestinal, renal, central or peripheral neurological involvement); a second model added ANCA positivity as an

input variable. Clinical relevance of the generated clusters was assessed by describing each cluster's clinical characteristics.

**Results:** We had data for 482 patients (46.3% males; mean age at diagnosis: 51 years [SD 14.9]) with a diagnosis mostly from 1990 to 2015. The cluster analysis generated 4 clusters of 34, 371, 39 and 38 patients. No distinctive cluster characteristic could be identified and we found no mutually exclusive subgroups, even after adding ANCA positivity to the model. ANCAs were detected in 37.1% of patients and were mostly P-ANCA (85.2%) or anti-myeloperoxidase (87.5%) according to the technique used. As compared with ANCA-negative EGPA patients, those with ANCA-positive EGPA were more often male ( $P = 0.01$ ), significantly younger ( $P = 0.002$ ), with more renal ( $P < 0.0001$ ) and peripheral neurological involvement ( $P < 0.0001$ ) and tissue findings of vasculitis ( $P = 0.01$ ), and fewer cardiovascular signs ( $P < 0.0001$ ) and tissue findings of eosinophilic infiltration ( $P < 0.0001$ ).

**Conclusion:** Although reinforcing the known associations of ANCA status with clinical manifestations, our data do not provide evidence for clearly distinct subclasses within EGPA. EGPA may be a phenotypic continuum rather than a phenotypic dichotomy, with entangled rather than distinct mechanistic pathways.

**Disclosures:** None