**Management and Outcomes of patients with ANCA-associated vasculitis (AAV) with pulmonary haemorrhage rescued with Extracorporeal Membrane Oxygenation (ECMO) – a series of 5 cases**

**Background**

ANCA-associated vasculitis (AAV) can manifest as pulmonary haemorrhage from small vessel inflammation of the lung capillaries. This can result in a spectrum of mild haemoptysis to life threatening diffuse alveolar haemorrhage requiring invasive respiratory support. In patients with active vasculitis the most common indication for admission to intensive care is respiratory failure or pulmonary renal syndrome. If pulmonary haemorrhage is so severe that gas exchange is inadequate despite maximal conventional ventilation strategies, ECMO may be used as a rescue therapy to oxygenate blood and remove carbon dioxide whilst allowing a reduction in potentially injurious ventilator pressures. We present a case series of five patients requiring ECMO support, their management, and outcomes.

**Aims**

To assess the management and outcome of pulmonary haemorrhage secondary to AAV in patients failing maximal conventional ventilation and requiring rescue ECMO in our centre.

**Methods**

We reviewed all the patients referred to our local ECMO centre in 2016 with a diagnosis of pulmonary AAV. We undertook a retrospective medical notes review of the five patients identified that failed optimal ventilatory management and required ECMO. We collected data on baseline characteristics, type of vasculitis, duration of ECMO, immunosuppressive therapies used, renal outcomes and patient survival.

**Results**

Five patients were identified as having diffuse alveolar haemorrhage requiring ECMO. Three patients were PR3-ANCA positive and two were ANCA negative. They required between 4 and 7 days of ECMO. All patients received methylprednisolone and between 5 and 7 plasma exchange treatments. Two received rituximab and three received intravenous cyclophosphamide for induction of remission. Three required renal replacement therapy but all achieved renal recovery. One patient died following further pulmonary haemorrhage after respiratory recovery and discontinuation of ECMO.

**Conclusions**

Lung involvement in AAV is common. In those failing conventional invasive mechanical ventilation, despite requirements for systemic anticoagulation ECMO would appear to be a valid rescue therapy with good overall outcomes. Both IV cyclophosphamide and IV Rituximab appear safe and effective in those patients requiring ECMO. Caution should be taken in those weaning off ECMO as late pulmonary haemorrhage may occur.