Apheresis treatment approaches for ANCA-associated vasculitis in children - synthesis of clinical data and case-based expert strategies

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Journal Name:

Pediatric Nephrology

Case vignette 1

Patient

12-year-old boy

Medical history and clinical presentation

Six-week history of fatigue, weight loss (-10%), and intermittent fever. Recurrent epistaxis and chronic rhinosinusitis for the past two years. No hemoptysis or dyspnea. Normotensive.

Laboratory findings

- Creatinine: 0.9 mg/dL (bedside Schwartz eGFR ~65 mL/min/1.73 m²)
- Urinalysis: protein-to-creatinine ratio 1.2 g/g, microscopic hematuria
- Hemoglobin: 9.8 g/dL
- CRP: 48 mg/L
- Strongly positive PR3-ANCA
- Anti-GBM antibodies negative

Kidney biopsy

Focal necrotizing pauci-immune glomerulonephritis with 25% crescents and no relevant fibrosis.

Questions related to case vignette 1

1. Which induction therapy would you primarily choose? (Single choice)

a. Glucocorticoids alone
b. Glucocorticoids + cyclophosphamide
c. Glucocorticoids + rituximab
d. Glucocorticoids + mycophenolate mofetil
e. Glucocorticoids + methotrexate
f. Glucocorticoids + cyclophosphamide + rituximab
g. Glucocorticoids + mycophenolate mofetil + rituximab
h. Glucocorticoids + methotrexate + rituximab
i. Other:

2. Would you use therapeutic apheresis? (Single choice)

a. Yes, directly as part of induction therapy.
b. Yes, if renal function deteriorates during follow-up.
c. Yes, if renal function and proteinuria do not improve within one month.
d. I would not use therapeutic apheresis.
e. Other:

3. Which form of therapeutic apheresis would you use? (Single choice)

a. Plasma exchange
b. Immunoadsorption

4. Please explain why you chose plasma exchange or immunoadsorption. (Single choice)

a. More scientific evidence
b. Greater institutional experience
c. Fewer side effects

d. Better reimbursement
e. Other:

5. Would you use off-label Avacopan? (Single choice)

a. Yes, directly during induction therapy.
b. Yes, in case of insufficient improvement.
c. Yes, in case of steroid toxicity.
d. No.
e. Other:

6. Which maintenance therapy would you prefer after remission induction? (Single choice)

a. Azathioprine
b. Mycophenolate mofetil
c. Methotrexate
d. Rituximab
e. Leflunomide
f. Other:

Case vignette 2

Patient

16-year-old female adolescent

Medical history and clinical presentation

Acute onset of disease 10 days prior with oliguria, edema, and arterial hypertension. No ENT manifestations and no pulmonary symptoms.

Laboratory findings

- Creatinine: 3.5 mg/dL (bedside Schwartz eGFR ~20 mL/min/1.73 m²)
- Urinalysis: protein-to-creatinine ratio 4.8 g/g, gross hematuria
- CRP: 92 mg/L
- MPO-ANCA positive
- Anti-GBM antibodies negative

Kidney biopsy

Diffuse necrotizing pauci-immune glomerulonephritis with >70% crescents and early interstitial fibrosis.

Questions related to case vignette 2

1. Which induction therapy would you primarily choose? (Single choice)

a. Glucocorticoids alone
b. Glucocorticoids + cyclophosphamide
c. Glucocorticoids + rituximab
d. Glucocorticoids + mycophenolate mofetil
e. Glucocorticoids + methotrexate
f. Glucocorticoids + cyclophosphamide + rituximab
g. Glucocorticoids + mycophenolate mofetil + rituximab

h. Glucocorticoids + methotrexate + rituximab
i. Other:

2. Would you use therapeutic apheresis? (Single choice)

a. Yes, directly as part of induction therapy.
b. Yes, if renal function deteriorates during follow-up.
c. Yes, if renal function and proteinuria do not improve within one month.
d. I would not use therapeutic apheresis.
e. Other:

3. Which form of therapeutic apheresis would you use? (Single choice)

a. Plasma exchange
b. Immunoadsorption

4. Please explain why you chose plasma exchange or immunoadsorption. (Single choice)

a. More scientific evidence
b. Greater institutional experience
c. Fewer side effects
d. Better reimbursement
e. Other:

5. Would you use off-label avacopan? (Single choice)

a. Yes, directly during induction therapy.
b. Yes, in case of insufficient improvement.
c. Yes, in case of steroid toxicity.
d. No.
e. Other:

6. Which maintenance therapy would you prefer after remission induction? (Single choice)

a. Azathioprine
b. Mycophenolate mofetil
c. Methotrexate
d. Rituximab
e. Leflunomide
f. Other:

Case vignette 3

Patient

17-year-old boy

Medical history and clinical presentation

Two-week history of progressive dyspnea, hemoptysis, and fever. Hospital admission due to respiratory failure. Arterial hypertension present.

Laboratory findings

- Creatinine: 2.8 mg/dL (bedside Schwartz eGFR ~30 mL/min/1.73 m²)
- Urinalysis: proteinuria 3.5 g/g, gross hematuria
- Hemoglobin: 7.9 g/dL

- CRP: 110 mg/L
- PR3-ANCA positive
- Anti-GBM antibodies negative

Imaging

Chest CT: Bilateral alveolar infiltrates consistent with alveolar hemorrhage.

Kidney biopsy

Necrotizing pauci-immune glomerulonephritis with 60% crescents and early interstitial fibrosis.

Questions related to case vignette 3

1. Which induction therapy would you primarily choose? (Single choice)

a. Glucocorticoids alone
b. Glucocorticoids + cyclophosphamide
c. Glucocorticoids + rituximab
d. Glucocorticoids + mycophenolate mofetil
e. Glucocorticoids + methotrexate
f. Glucocorticoids + cyclophosphamide + rituximab
g. Glucocorticoids + mycophenolate mofetil + rituximab
h. Glucocorticoids + methotrexate + rituximab
i. Other:

2. Would you use therapeutic apheresis? (Single choice)

a. Yes, directly as part of induction therapy.
b. Yes, if renal function deteriorates during follow-up.
c. Yes, if renal function and proteinuria do not improve within one month.
d. I would not use therapeutic apheresis.
e. Other:

3. Which form of therapeutic apheresis would you use? (Single choice)

a. Plasma exchange
b. Immunoadsorption

4. Please explain why you chose plasma exchange or immunoadsorption. (Single choice)

a. More scientific evidence
b. Greater institutional experience
c. Fewer side effects
d. Better reimbursement
e. Other:

5. Would you use off-label avacopan? (Single choice)

a. Yes, directly during induction therapy.
b. Yes, in case of insufficient improvement.
c. Yes, in case of steroid toxicity.
d. No.
e. Other:

6. Which maintenance therapy would you prefer after remission induction? (Single choice)

a. Azathioprine
b. Mycophenolate mofetil

c. Methotrexate
d. Rituximab
e. Leflunomide
f. Other: