Hypercortisolism Management

Short-term management

- Metyrapone
  - First-line due to low hepatotoxicity
  - Starting dose: 300 mg/m²/day divided BID
  - Usual blocking dose: 600 mg/m²/day
  - Max dose: 1200 mg/m²/day

- Etomidate
  - For critically ill patients unstable for surgery
  - Use in addition to metyrapone
  - Starting dose: 0.25 mg
  - Usual blocking dose: 1 mg/kg

- Other medical therapies
  - Mitotane, ketoconazole
  - Little experience with use in MAS
  - Use with caution due to hepatotoxicity

Long-term management

- Adrenalectomy
  - Bilateral usually required (may consider unilateral if disease limited to one side on CT)
  - Spontaneous resolution seen in approx. one third—consider watchful waiting in patients with mild disease
  - Monitor cognitive development

1Patients are often critically ill at presentation, which may impact treatment options. 2Hepatotoxicity is an important consideration due to frequent comorbid liver disease. 3Spontaneous resolution may occur due to involution of the adrenal fetal zone, which is the source of hypercortisolism in MAS. 4Children with a current or remote history of MAS-associated hypercortisolism are at increased risk for neurodevelopmental delays, and should be considered for early interventional services.

References

Legend
BID = twice daily; CT = computed tomography; m² = meters squared; MAS = McCune-Albright syndrome