

CONVENTIONAL TREATMENT OF INFECTION/INFLAMMATION IN CHRONIC GRANULOMATOUS DISEASE (CGD)

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CGD is a complex immune deficiency with both infection susceptibility and dysregulated inflammation pointing to two protective NADPH oxidase functions: Inactivation of pathogens and resolution of inflammation. Balanced pharmacologic targeting of the two pathways is the basis of CGD management and has resulted in new approaches of conventional treatment.

Currently glucocorticoids are used empirically as main anti-inflammatory agents in CGD. Their cautious use is best illustrated in three conditions:

Liver abscesses are mostly due to *S. aureus*. The septated, pyogranulomatous lesions once required invasive surgical excision. Today surgery is best avoided and replaced by prednisone 1mg/kg/day 2-3 wks (plus antibiotic therapy).

1. Fulminant miliary pneumonitis by massive inhalation of *Aspergillus* spores results in miliary inflammatory infiltrates requiring ventilation. An iv combination of antifungals AND steroids is life-saving.
2. „Sterile“ granulomatous colitis mimicking Crohn's disease is again responsive to prednisone 1 mg/kg/day 1-2 wks, slow taper.

Immunosuppressed CGD patients must be covered by antimicrobials. Ongoing molecular dissection of antiinflammatory pathways (e.g. of PPAR gamma activation) will yield novel interventions avoiding many of the steroid side effects in the near future.