CGDSociety
Supporting families affected by chronic granulomatous disorder

The Chronic Granulomatous Disorder Society
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Dear Clinician,

I am/my child is affected by the rare, genetic condition Chronic Granulomatous Disorder (CGD) (also known as Chronic Granulomatous Disease). The condition affects the way the immune system fights infection.

This letter aims to help you understand the condition and the need for prompt and specialist treatment if I/my child is unwell away from home. You can find more information at www.cgdsociety.org.

I have the details of the doctors who look after my/my child's CGD in my home country. I also have a contact name of a doctor in this country that can provide you with help and advice if needed.

What is CGD?
The condition develops when phagocytic cells (white blood cells, including neutrophils and monocytes) fail to effectively destroy invading bacteria and fungi. This makes me/my child susceptible to serious, potentially life-threatening, bacterial and fungal infections. I/my child can also have symptoms associated with chronic inflammation, often granulomatous in nature.

What symptoms need prompt further investigation and treatment?
Symptoms that need to be assessed and treated urgently include:
• Persistent diarrhoea (travellers' diarrhoea may have very serious implications for my health)
• A fever of 38ºC or above
• Infected cuts and wounds
• Persistent cough or chest pain
• Acute shortness of breath/shortness of oxygen to the tissues or body after exposure to organic matter (e.g. leaves, compost)
• Warm, tender or swollen areas, lumps, boils or significant rashes
• Pain or difficulty urinating
• Vomiting shortly after eating (on a more or less consistent basis) or difficulty swallowing food

Advise on treatment of acute infection:
Any illness characterised by fever should be treated QUICKLY with antibiotics, followed by appropriate intravenous therapy if needed. A 'safety-first' approach should always be adopted for patients with CGD regardless of concern over inappropriate treatment of viral infections/overuse of antibiotics. Patients with CGD may require longer courses of antibiotics, sometimes at higher doses, or combination of antibiotics, because of their poor immunity.

If a patient's symptoms do not improve to initial treatment, we strongly recommend you seek advice from a specialist. They may suggest that empirical anti-fungal treatment is indicated.

Oral antibiotics:
Oral Ciprofloxacin is a good first line agent because of its spectrum of activity and capacity to penetrate intracellularly. The benefits of using Ciprofloxacin in children with CGD outweigh the risks of developing joint disease (arthropathy).

DOSE: Child: 7.5 mg/kg; every 12 hours
Intravenous antibiotics:
IV Teicoplanin and Ciprofloxacin are a good choice for initial treatment of severe sepsis. Metronidazole can be added if infection below the diaphragm is suspected.

Antibiotics should be modified in the light of any positive cultures, but combinations or prolonged courses are advised for deep seated infections e.g. Flucloxacillin plus clindamycin for staphylococcal infection.

DOSE: Adult:
Ciprofloxacin (oral or IV) 500mg to 750mg every 12 hours
Teicoplanin 10mg/kg every 12 hours for first 3 doses then every 24 hours
Metronidazole 400mg (oral) or 500mg (IV) every 8 hours

An alternative to the above combination is meropenem 1g every 8 hours, if available, or another broad-spectrum carbapenem (e.g. Iimipenem).

This information is endorsed by the CGD Society Medical Advisory Panel:
http://www.cgdsociety.org/about/medicaladvisorypanel

Thank you for reading this information.

The Chronic Granulomatous Disorder (CGD) Society