Chronic Granulomatous Disease, Osteopetrosis and Actimmune® (interferon gamma-1b)

What is Chronic Granulomatous Disease (CGD) and Osteopetrosis?

Table 1 provides a general overview of CGD and osteopetrosis.

Table 1: CGD and Osteopetrosis

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| Chronic Granulomatous Disease (CGD) | A rare disease caused by a defect in the immune system that makes it hard for the body to fight off certain bacterial and fungal infections | People who have an abnormal gene that is passed down from a parent or parents, boys are more likely to have CGD than girls; occurs in approximately 1 out of every 1 million people | • Frequent bacterial and fungal infections, including pneumonia and skin or bone infections  
• Swollen lymph nodes and tissues (also called granulomas) |
| Osteopetrosis                      | A rare disorder that causes the bone to become overly dense. People with osteopetrosis have bones that are more brittle than normal. This happens because your body can’t replace old bone with new bone at a normal speed. A mild form of osteopetrosis may cause no problems, However, in severe forms, the overgrown bone decreases bone marrow activity and it can also cause stunted growth, deformity and | Children who have an abnormal gene that is passed down from parents | • Blindness  
• Hearing loss  
• Unable to control facial muscles  
• Anemia  
• Frequent infections and bone fractures |
How can I stay healthy?

For people with Chronic Granulomatous Disease

- Careful personal hygiene
  - Brush your teeth two times daily with hydrogen peroxide and baking soda paste to avoid gum disease and canker sores.
  - Wash cuts right away and clean with an antiseptic.

- Avoid alcohol and tobacco use

- Make sure you are up-to-date with your immunizations

- Lower your chance of getting an infection
  - Don’t swim in rivers or lakes, even if they look clean. They can be full of bacteria.
  - Choose playgrounds with plain dirt or gravel surface – wood chippings may have a lot of fungi.
  - If you use a humidifier, empty it daily and wash it with bleach to prevent mold.

- Contact your doctor right away if you have a fever with a cough.

For people with osteopetrosis

- Good nutrition
  - Eat a balanced diet
  - This will help support normal growth and development.

- Frequent ear, nose, and throat check ups
  - An otolaryngologist (ear, nose, and throat doctor) should check blocked nose and throat airways often.

- Orthopedic Care and Fracture Prevention
  - If you have frequent bone fracture, make sure to follow up with your orthopedic doctor.

What is Actimmune (interferon gamma-1b)?

- Actimmune (interferon gamma-1b) is a medication that acts like a substance in your body called interferon gamma.
- It reduces the number of serious infections from chronic granulomatous disease (CGD).
- It may help to slow down the progression of severe, malignant osteopetrosis.
- This medication may have other uses; ask your doctor for more information.
**How do I take Actimmune?**

- Do not use Actimmune until your doctor has trained you in the proper injection techniques.
- It is injected under the skin three times weekly (for example, Monday, Wednesday, Friday).
- Inject in the upper arms, abdomen, or thighs.
- It is important to change the site of an injection each time you give the medication.

**How should I store Actimmune?**

- Actimmune vials must be stored in the refrigerator, 2-8° C (36-46°F).
- Do not freeze.
- Do not shake.
- Throw away any unused vials if they have been out of the refrigerator longer than 12 hours.
- Do not use any vials after the expiration date.

**What can I do to make side effects tolerable?**

- The most common side effects with Actimmune are “flu-like” symptoms such as fever, headache, chills, muscle ache, or fatigue.
- These symptoms may get better as you continue with your medication.
- You can reduce “flu-like” symptoms by injecting the medication at bedtime or in the evening. You may sleep through the symptoms if you take it early in the evening.
- Ask your doctor if you can use over-the-counter medications (such as Advil®, Motrin®, or Tylenol®) for fever and headache.
- Change the site of an injection each time you give the medication to reduce skin irritation and redness.
- Contact your doctor right away if you have any of the following problems at the injection site:
  - Lumps, swelling, or bruising that doesn’t go away
  - Any signs of infections or swelling such as pus, redness, skin surrounding the injection site that is hot to touch, and pain after the injection.
- If symptoms continue or you have any questions or concerns, please talk to your doctor.
What other resources are available?
For more information, please contact the following resources:

- **OptumRx**
  - Phone: **1-800-562-6223**
  - Website: www.optumrx.com

- **Chronic Granulomatous Disease**
  - NIH/National Institute of Allergy and Infectious Disease (NIAID)
    - Phone: 866-294-4107
    - Website: www.niaid.nih.gov
  - National Association for Rare Disorders (NORD)
    - Phone: 800-999-6673
    - Website: www.rarediseases.org
  - Chronic Granulomatous Disease Association, Inc
    - Phone: 626-441-4118
    - Website: www.cgdassociation.org
  - Immune Deficiency Foundation
    - Phone: 800-296-4433
    - Website: www.primaryimmune.org

- **Osteopetrosis**
  - National Association for Rare Disorders (NORD)
    - Phone: 203-711-0100
    - Website: www.rarediseases.org
  - The Paget Foundation
    - Phone: 800-237-2438
    - Website: www.paget.org
References:

The information in this educational handout doesn’t substitute the medical advice, diagnosis, or treatment provided by your doctor. Always seek the help of your doctor or a qualified health provider for any questions about your medical condition.