Orofacial Granulomatosis

What is orofacial granulomatosis?
Orofacial granulomatosis (OFG) is an uncommon condition of the mouth that causes lip swelling, and sometimes swelling of the face, inner cheeks, and the gums. This swelling may come and go at first, but over time, becomes persistent if not treated. Patients will typically describe their symptoms as swelling and tightness and sometimes, soreness. In addition, there may be some folding of the inside of the mouth giving it a “cobblestone” appearance. OFG may be part of Melkersson-Rosenthal syndrome, a rare condition that consists of OFG, fissured tongue (tongue with deep grooves) and paralysis of the face.

What causes OFG?
It is likely that patients with OFG are reacting to something although this is not always easy to identify. Some common triggers are foods and food preservatives such as chocolate, cinnamon flavorings, and preservatives such as benzoates and metabisulphites. In some studies, one-third of patients will show great improvement just by adhering to a strict diet that has few or no preservatives, and by eliminating as much as possible, processed foods. In addition to the items mentioned above, other foods that have been associated with this condition include dairy products, wheat products and eggs.

In some cases OFG may be associated with Crohn disease, which is a disease of the gastrointestinal (digestive) tract that can cause diarrhea, abdominal cramping and bloody stools. If you developed OFG when you were a child, you are somewhat more likely to develop Crohn disease as you get older. In addition to lip/mouth swelling and folding of the mouth tissues, patients with Crohn disease may have mouth ulcers similar to canker sores.

How do we know it is OFG?
A dental specialist can usually diagnose OFG based on a good history, a thorough clinical examination, and a biopsy to confirm the diagnosis. You may be sent to a specialist who can perform a patch test of your skin to see if you are sensitive to foods, food preservatives or other substances. The results of the patch test may help your doctor to suggest the best treatment option for your disease. Patients may be referred to a gastrointestinal specialist if a diagnosis of Crohn disease is suspected.

How do we treat OFG?
A food diary can be helpful if food sensitivity is suspected or confirmed by patch testing, and completely avoiding such foods may substantially reduce swelling. The doctor who patch tested you will provide you with a list of substances to avoid if you patch tested positive to them. However, in other cases, there is no trigger, or it is very difficult to completely eliminate the trigger (such as food preservatives that are widely used). In such cases, the goal is to control the disease by reducing the amount of inflammation, thereby reducing swelling and pain. How it is treated depends on the severity of the condition.

You will likely be treated with topical steroids 3-4 times a day for a few weeks. The most commonly prescribed topical steroids are fluocinonide or clobetasol gel, and dexamethasone solution. Sometimes your doctor may recommend treating the area “intralesionally” (with a steroid injection directly into the involved area), to speed the healing process. In severe cases, steroid tablets such as prednisone may need to be taken for several weeks to reduce swelling. This may need to be done now and then during flare-ups. You may also be given other medications by mouth for long-term treatment and your doctor will discuss these with you. In cases of OFG associated with Crohn disease, you will likely be treated by your GI doctor with a medication for Crohn disease which may also be effective in controlling the OFG. Otherwise, topical and intralesional steroids will be used as additional treatments.
What can I expect?
In some patients, elimination of an identifiable triggering agent will cause the OFG to go away. However, most cases of OFG run a chronic course and may take many months or even years to resolve, even with treatment. Some patients have to be on long-term medications by mouth to control swelling and discomfort. If you begin to experience gastrointestinal symptoms further medical attention is necessary to assess for possible Crohn disease.