GI Medications

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Gastrointestinal problems are common in Cornelia de Lange Syndrome (CdLS) and are often treated with a confusing array of medications. I have broken some of the commonly used medications down by classification to discuss their indications and special considerations.

Medications that block stomach acid production: These medications are used when stomach acid is irritating the esophagus during gastroesophageal reflux. They can also be used when there is acid irritation to the stomach or when stomach acid is leaking out a G-tube site. These medications fall into two categories, H2 blockers and Proton Pump Inhibitors. They produce the same result by different mechanisms.

H2 blockers: This class of medications include Famotidine, Cimetidine, Ranitidine, and Nizatidine. They are usually given in two or more doses a day and come in liquid and pill formulations. They are user friendly in that they work well when given with or without food. They may be given in combination with Proton Pump Inhibitors, especially at night.

Proton Pump Inhibitors: This group of medications is more potent than the H2 blockers and include Omeprazole, Lansoprazole, Zegrid, Rabeprazole, Pantoprazole, and Esomeprazole. These medications are not as user friendly in that they don’t all come in liquid preparations or preparations meant to be opened and mixed in food. They are more effective when given without food. There is concern that long term use may lead to osteoporosis, making fractures more likely.

Medications for constipation: Constipation is a common problem in CdLS. Commonly used and effective medications include osmotic medications, stimulants and rectally administered medications.

Osmotic medications hold water in the stool making it easier to pass. This prevents hard stools that are painful to pass. There are several formulations which can be mixed with any fluid. They may be taken by mouth or administered down a feeding tube. They do not increase gas production. Examples include Miralax, Colyte, Glycolax, GoLytely, NuLytely, and Trilyte.

Milk of Magnesia is also an osmotic agent for constipation. Care should be given to not administer it with other medications because it may bind them and change drug levels.
**Stimulant laxatives:** These laxatives increase the muscle contractions in the colon and may increase the urge to have a bowel movement. They are often used in combination with osmotic agents in special needs patients. Senna is a mild stimulant and comes in many forms including liquid, chocolate chews and pills. It can safely be used for long periods of time. Bisacodyl is a more potent stimulant laxative. It comes in the form of small pills which can not be crushed. It also comes as a suppository which can be used every few days to stimulate a bowel movement. It can also be used as a chronic medication.

**Rectal medications:** A variety of medications for constipation can be given into the rectum.

Mineral oil can be used as an enema to soften hard stools. In general it should not be given by mouth or feeding tube in CdLS because of the chance of aspiration pneumonia.

Saline can also be used as an enema to help pass stools. It can be mixed with one teaspoon of salt for four cups of tap water.

Phosphate enemas should be avoided in CdLS because it may cause severe problems with blood salts.

Glycerin suppositories can provide mild stimulation to have a bowel movement. Bisacodyl suppositories are more potent. Both can be used on a regular basis.