MEDICATIONS/DIAGNOSTICS

Individuals with Dent Disease need to be cautious with certain medications and diagnostic imaging. Due to possible nephrotoxic effects please be aware that the following should be assessed and prioritized prior to treatment in a hospital/clinic setting:

- If possible, avoid NSAIDS
- Double check antibiotics for nephrotoxic adverse effects
- Contrast dyes used in CT scans, angiograms, and MRI’s should also be prioritized due to possible nephrotoxic effects.

*Medical consultation is advised for best outcome.*

ACUTE MANAGEMENT OF DENT DISEASE

Dent disease patients are at risk for dehydration and should be medically treated to avoid this. Close monitoring of electrolytes is also important. If kidney stones are present, preventing blockages in the ureters and maintaining kidney function is priority.

PREOPERATIVE EVALUATION AND SURGERY

A preoperative evaluation of kidney function should be performed on all Dent disease patients regardless of age and whether or not they have chronic kidney disease. Contact the patient’s Nephrologist if possible before surgery as they are aware of special needs required for best outcome.

FOR MORE INFORMATION PHYSICIANS CAN LOG INTO WWW.UPTODATE.COM OR CONTACT:

www.dentdisease.org

QUICK INFORMATION FOR THE CARE OF PATIENTS WITH DENT DISEASE

**Name:**
**Date of Birth:**
**Name/Relationship of caregiver if patient is a minor:**

**NEPHROLOGIST**
**Name:**
**Phone:**
**Email:**

**EMERGENCY CONTACT:**
**Name:**
**Phone:**
**Phone:**

**Dent disease is a chronic kidney disease that primarily affects males.** While symptoms and severity vary, they usually begin in childhood and worsen over time. The most common feature of Dent disease is proteinuria (protein in the urine). Other common features include excess calcium in the urine (hypercalciuria); calcium deposits in the kidneys (nephrocalcinosis); and kidney stones. Less common features include rickets and mildly short stature. Progressive kidney problems often lead to kidney failure by early to mid-adulthood.

There are two forms of Dent disease which are distinguished based on their genetic causes. Both forms are inherited in an X-linked recessive manner.

**TYPE 1**
Caused by a mutation in the CLCN5 gene

**TYPE 2**
Caused by a mutation in the OCRL gene

_Males with this form are also at increased risk for mild intellectual disability and hypotonia._
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