Medullary sponge kidney

Introduction
• **Definition:** A dilation of the distal convoluted tubules in the pericalyceal area of the renal pyramid. Associated with medullary cysts. May be unilateral or bilateral.
• **Other names:** *Sponge* kidney or *Cacchi-Ricci* disease.
• **Heredity:** Although there is no evidence of genetic transmission, some families seem to show autosomal dominant inheritance.
• **Prevalence:** <1% of urographic studies.
• **Associated with phosphate and calcium oxalate lithiasis.**
• **Risk factors:** Urinary stasis, increased urinary pH, hypercalciuria, hyperuricosuria, and hypocitraturia all contribute to the intraductal deposit of crystals.

Symptoms
• **Asymptomatic.** In most patients it is a urographic finding.
• **If associated with nephrocalcinosis,** patient usually presents *low back pain* and *hematuria.*
• **May be associated with hypercalciuria and hypocitraturia.**
• **UI** present in 1/3 of patients.

Diagnosis
• **IVP:** The ideal method. The images show:
  - *Brush border* at the edge of the stones due to tubular dilation.
  - Increased size of the pyramids.
  - *Clustered* intraductal microcalcifications.
• **Ultrasound:** Corticomedullary junction is uniformly echogenic due to calcium deposits.
• **CT scan:** Although conventional CT are less sensitive than IVP in diagnosing tubular ectasia, new techniques of helical CT and uro-CT have improved its efficacy and show distal renal tubules filled with contrast medium.

Prognosis
• **While medium and long term prognosis is very good,** in some cases the associated stones and UTI can lead to chronic renal failure.

Treatment
• **Observation:** In cases of incidental discovery in asymptomatic patients.
• **In cases of symptomatic lithiasis:** After the migration of the stone to the ureter, conventional treatments such as *ESWL* and ureteroscopy are used.
• **Drug treatment** to alleviate associated alterations such as hypercalciuria and hypocitraturia.

<table>
<thead>
<tr>
<th>Generic name</th>
<th>Brand name</th>
<th>Dose</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Potassium citrate*</td>
<td>ACALKA, BLANEL</td>
<td>1 tab of 1.08 g/8 h</td>
<td>months</td>
</tr>
<tr>
<td>Potassium citrate-citric ac</td>
<td>URALYT URATO, RENAPUR</td>
<td>2.5 g (1 teaspoon)/8 h</td>
<td>months</td>
</tr>
<tr>
<td>Sodium bicarbonate</td>
<td>SODIUM BICARBONATE</td>
<td>2 tab of 500 mg/6-8 h</td>
<td>months</td>
</tr>
<tr>
<td>Hydrochlorothiazide**</td>
<td>MODURETIC</td>
<td>1 tab of 50 mg/d</td>
<td>months</td>
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</tbody>
</table>

*After meals. In case of renal insufficiency, Sodium bicarbonate (no potassium) is preferred.*

**Associated with 5 mg of Amiloride in order to reduce hypokalemia.**