Guidelines for management of a child with renal stone or nephrocalcinosis

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These guidelines were produced in good faith by the author(s) reviewing available evidence/opinion. They were designed for use by paediatric nephrologists at the University Hospital of Wales, Cardiff for children under their care. They are neither policies nor protocols but are intended to serve only as guidelines. They are not intended to replace clinical judgment or dictate care of individual patients. Responsibility and decision-making (including checking drug doses) for a specific patient lie with the physician and staff caring for that particular patient.

Version 1/V Bisht and S Hegde /Nov 2009

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Introduction

Stones in kidneys or urinary tract are not uncommon in children. *Nephrolithiasis* (kidney stones) or *nephrocalcinosis* (increased calcium content in the kidneys, which may be focal or generalised) are often a sign of underlying disease/anatomical anomalies and not the disease itself. Stones can be infective or metabolic or combined. They are more common in boys. Most stones affect the upper tract. Contrary to earlier belief that stones in children are mostly infection related, recent epidemiological data suggests majority having underlying metabolic abnormality. Diagnostic evaluation in every child (even with a single stone) is necessary to prevent recurrence and adverse outcomes like renal failure.

History and physical examination

Classic symptoms of **stones** (e.g. renal colic and haematuria) are *uncommon* in children. More commonly they present with abdominal pain, vomiting, fever, signs of UTI, micro or macroscopic haematuria. Small children often present with non specific signs. Some children may have sterile pyuria, dysuria, voiding problems, urine retention, enuresis, frequent voiding or history of passage of stones or gravel. Some stones are detected incidentally.

Nephrocalcinosis is mostly asymptomatic and often detected incidentally. They might rarely present with renal colic (passage of tiny calculi), haematuria (gross or microscopic) or sterile leukocyturia.

It is important to obtain information on-

- Family history of stones, metabolic disease and renal failure
- Diet history – high in animal protein/ ketogenic diet for epilepsy, excessive salt intake
- Poor fluid intake, excessive intake of fizzy drinks
- Medications- vitamin D/A, steroids, diuretics, anticonvulsants, chemotherapy, ceftriaxone
- Prolonged immobilisation
- Mineral supplements i.e. calcium
- Chronic bowel disease- Crohn's disease, bowel resection, chronic diarrhoea
- Urinary tract anomalies (leading to stasis and UTI)- neurogenic bladder, ileal loops , megaureter/megacalycosis, hydronephrosis
- **Cystic Fibrosis** – high risk of calcium stone formation due to combination of low urine output, hyperoxaluria and hypocitraturia.
- **Inherited disorders**- Bartter's, Lowe's and Williams syndrome, Dent's disease, Hyperoxaluria, cystinuria, autosomal dominant hypocalcemic hypercalciuria and familial hypomagnesemia with hypercalciuria and nephrocalcinosis (FHHNC), RTA, inborn errors of purine metabolism, xanthinuria

Physical examination should include evaluation of growth and development, bone structure and blood pressure.

Classification of renal stones

Infection related stones are composed of struvite (magnesium ammonium phosphate and calcium carbonate-apatite) and are caused by urease producing organism i.e proteus. However various metabolic disorders given below may lead to formation of stones composed of calcium oxalate, calcium phosphate, uric acid and cystine.

Struvite stones (magnesium ammonium phosphate and calcium carbonate-apatite)	Calcium stones	Uric acid stones	Oxalate stones	Rare stones	Other
Associated with UTIs	Disorders of hypercalciuria & hypercalcemia (see guidelines) Dent's disease	Inborn errors of purine metabolism Tumour lysis Chronic diarrhoea	Hyperoxaluria (primary, enteric or idiopathic)	Xanthinuria Orotic aciduria	Cystine stones in cystinuria Stones in Hypocitraturia
Radio-opaque	*Radio-opaque*	*Radio-lucent*	*Radio-opaque*	*Radio-lucent*	*Radio-opaque*

Investigations

Blood- U&Es, bicarbonate, venous gas, chloride, liver functions, bone profile, magnesium, urate, PTH, plasma oxalate.
Selected cases- Vitamin A & D levels, plasma oxalate & vitamin B6 levels

Urine - for urine samples child should be on normal diet and fluid intake. Avoid collections when on i/v fluids or post operative or when being treated for UTI. A properly collected 24hr collection (need acidified containers) provides most reliable data but spot samples are usually used because of ease of collection. Spot samples (usually a *second* morning urine sample) can be collected in normal containers but need to be sent to lab straight away (marked as "*for attention of special chemistry*"). Contact lab at ext 8364 prior to sending specimen.

Urine should be sent for -

- Dipstick, microscopy (RBC morphology & crystals e.g. cystine) and culture
- Oxalate: creatinine ratio
- Protein: creatinine ratio
- PH (fresh sample to lab, *not by dipstick*)
- Calcium: creatinine ratio
- Urate: creatinine ratio
- Cystine: creatinine ratio
- Citrate: creatinine ratio
- Aminoacid screen

Also- Magnesium, osmolality, electrolytes

If hyperoxaluria present send further samples for urine **glycolate: creatinine ratio** (PH Type I) and analyse urine for **glyceric acid** by organic acid screen (PH Type II).

Abnormal parameters are confirmed on 2-3 occasions. Try and confirm abnormal results with a 24 hour specimen (ensuring normal fluid intake and normal dietary habits).

Radiology

- US KUB
- (Plain x-ray)
- CT scan if small/suspected ureteric stones
- MAG 3 if suspected obstruction

Stone analysis
If stone passed or retrieved, should be sent for complete quantitative analysis (very useful).

Management

If suspected obstruction due to stone, hospitalise, urgent referral to urologist, check kidney functions, may require urgent intervention.

If no obstruction, proceed for metabolic work up, consult urologist and nephrologist. Manage conservatively with adequate fluid intake and analgesia. Treat underlying metabolic disorder. If stone does not pass spontaneously definitive treatment is needed in the form of extracorporeal shock wave lithotripsy (ESWL), percutaneous nephrolithotomy or endoscopic removal depending on size, location and type of stone.

General recommendations

- Increased fluid intake (minimum125% of normal maintenance volume) and avoidance of fizzy drinks
- Decreased intake of animal protein
- Restricted salt intake
- Normal calcium intake (not in excess of or less than recommended amount)
- Reduce intake of food rich in oxalate
- Treatment of underlying disorder (e.g. cystic fibrosis)
- Potassium citrate- might be helpful even in the absence of hypocitraturia

Specific recommendations

For calcium stones – oral potassium citrate, thiazide diuretics to reduce hypercalciuria
Uric acid stones- allopurinol, potassium citrate
Oxalate stones- no specific drug. Consider vit B6
Cystine stones- alkanisation with potassium citrate, consider specific agents like mercaptopropionylglycine or d-penicillamine.
Struvite stones – prompt treatment of infection.

References

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Management algorithm for renal stone disease