Rickets, diagnosis and management

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25th May 2013
Rickets

A disease of deficient mineralization at the level of growth plate and is usually accompanied by osteomalacia i.e., impaired mineralization of bone matrix.

Rickets and osteomalacia occur together as long as the growth plate is open, only osteomalacia when growth plate is closed.

*PittMj Radiol Clin North America 1991;29:97*

It affects infants, children, and adolescents.
Rickets

Undermineralised bones are soft and can be bent and twisted under the continuing influence of gravity or even opposing muscle tension.

The bones are at risk of being deformed at the growing ends of long bones, risk of fractures.
Rickets

Results from lack of 2 essential substances

1. Calcium

2. Phosphate

*J Clin Endocrinol Metab 2003; 88:3539-45*
RICKETS

Hypocalcaemic
- Vit D deficiency
- Defective metabolism
  - Vit D production: vit D dependant dependant
  - Vit D resistance :vit D resistance

Hypophosphophaemic
Causes of Rickets

**Calciopenic**

- Nutritional
  - Vit D deficiency
  - Calcium deficiency
    - (soy based, phytate)

- Inherited
  - $\alpha$ hydroxylase deficiency
    - (Vit D dependant rickets Type 1)
    - Resistance to 1,25 (OH)$_2$ VitD
      - (Vit D dependant rickets type 2)

**Phosphopenic**

- Phosphate deficiency
  - (prematurity)

- Inherited
  - Hypophosphataemic rickets
    - (x linked, autosomal dominant)
  - Hereditary hypophosphatemic rickets
    - with hypercalciuria (HHRH)
      - (x linked, autosomal dominant)
Rickets

**calciopenic**

- Malabsorption vit D
  - IBD
  - short bowel syndrome
  - Cystic fibrosis
  - Coeliac dis

- Impaired hydroxylation of Vit D
  - severe hepatobiliary dis
  - severe renal dis

- Increased catabolism of Vit D

**phosphopenic**

- Impaired absorption of phosphate
  - severe intestinal dis
  - Phosphate binding gels
    - (Aluminum containing antacids)

- Phosphate wasting renal tubulopathy

- Tumour induced rickets

- Fanconi syndrome
Prevalence of rickets

Vitamin D deficiency is the commonest cause of rickets worldwide

*J Clin Nutr* 2008;87(S1):1080S-6S
Main source of Vit D for humans is from exposure to sunlight (UVB 290-315nm)

*J Cell Biochem 2003; 88: 296-307*

*J Clin Invest 2006:116:2062-2072*
Sunshine main source
30 min / week (diaper)
2 hr / week fully clothed

D3 cholecalciferol (sun)
D2 ergo calciferol (Food)
Nomenclature of Vit D

Forms of Vit D

Vit D2 (food) (ergocalciferol)
Vit D3 (cholecalciferol)
1α hydroxy vit D (α calcidiol)
1,25 dihydroxy vit D (rocalcitriol)
VIT D deficiency

Infants younger than 6 months confined indoors

Infants dependant on food they consumed (milk before weaning 6 months)

Breast milk poor source of vit D (12-60IU/L)

*J Nutr* 1981;111:1240-1248

*J Ped* 1982;100:745-748
Vit D deficiency

The main risk factors
barriers to sun exposure
dark skin pigmentation
exclusive breast feeding
disorders of gut, liver, kidney

Ann Trop Paed 2006; 26(1): 1-16
Endocrinol Metab Clin North Am 2005; 34(3) : 537-553
Diagnosis

Clinical presentation variable

Neonatal and Infancy
hypocalcaemic fits
Tetany
apnoe and stridor
muscle weakness
delayed motor milestones/ hypotonia
soft skull bones, wide open sutures
Clinical presentation

hypocalcaemic cardiac myopathy
dilated /hypertrophied cardiomyopathy
prolonged QTc interval, arrhythmias
hypotension and cardiac failure
Clinical presentation

OTHER SYMPTOMS & SIGNS

• Weakness, leg pain during walking
• Excessive falling
• Leg pain at rest
• Unable to walk
• Previous fracture
• Delayed dentition
• Skeletal deformities (bow legs/knock knees)
• Short stature
Clinical signs of Rickets
Radiological changes

Advance through defined stages

Loss of demarcation bet epiphysis & metaphysis
Metaphysis cupping, fraying, splaying

Healing begins a thin white line of calcification at junction of growth plate & metaphyseal area

Periosteum appears separated from diaphysis due to a layer of unmineralised osteoid

Gen osteopaenia with visible coarsening of trabeculae due to sec hyperparathyroidism
Radiological changes
Diagnostic approach

Question

is this hypocalcaemic
or
hypohosphataemic rickets?
Biochemistry

First

evaluate the serum calcium, phosphate and alkaline phosphate
Pitfalls

If you are not aware that normal reference range for phosphate varies with age that it is highest in neonates than in older child and adult

You will mistakenly regard it as normal when you use the adult reference range

underdiagnosis of hypophosphatemic rickets in prem and neonates
Phosphate

Values vary in relation to age and growth

PO₄ mmol/L

Prem
Newborn
Infant <6/12
Child 1 year
Childhood
Adult

2.6
1.6-2.5
1.3-2.4
1.3-2.1
1.2-1.9
0.6-1.5
Pearls

Always take fasting sample, as food affects Phosphate values causing falsely high $\text{PO}_4$

In hypocalcemic rickets PTH will respond to normalize the plasma calcium by acting at 3 sites
Should Know PTH effects

**Parathyroid hormone actions**

- **Serum**
  - Calcium
  - Phosphate, HCO₃
  - ALP, chloride
  - I :25 (OH)Vit D
  - Calcium reabsorption

- **Urine**
  - HCO₃ loss, PO₄
  - aminoaciduria

**NOTE:**
Secondary hyperparathyroidism due to hypocalcaemia can be mistaken as RTA.
Pitfall

In hypocalcaemic rickets the serum calcium levels is not always low, it can be normal depending on the stage of disease.
Remember: 3 stages in devt of hypocalcaemic rickets

1\textsuperscript{st} stage: hypocalcaemia (early phase)
- low Calcium
- a rise of PTH

2\textsuperscript{nd} stage: hypocalacemia continue to fall, PTH rises
- further normalising calcium at expense of PO4,
- ALP high and 1,25(OH)D maintained or high

3\textsuperscript{rd} stage: further fall of calcium, but because PTH is maintained at high level to raise calcium to normal value
- effect of hyperparathyroidism is seen (radiologically) and biochemically with hypophosphatemia, raised ALP, HCO3 wastage, aminoaciduria
- clinical: Rickets severe at this stage
Phases in response to low calcium: interpretation of results

<table>
<thead>
<tr>
<th></th>
<th>1&lt;sup&gt;st&lt;/sup&gt; phase Low calcium Normal P0&lt;sub&gt;4&lt;/sub&gt;</th>
<th>2&lt;sup&gt;nd&lt;/sup&gt; phase Low /normal Ca</th>
<th>3&lt;sup&gt;rd&lt;/sup&gt; phase Normal Ca Low P0&lt;sub&gt;4&lt;/sub&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calcium</td>
<td>normal low</td>
<td></td>
<td></td>
</tr>
<tr>
<td>phosphate</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PTH</td>
<td>N or</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ALP</td>
<td>N or</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
You might mistake hypocalcaemic rickets as hyposphosphatemic rickets or RTA.

Thus always order PTH so that you can make the correct interpretation at the initial diagnosis.
Pitfall

Calcium 2.2 - 2.6 mmol/L

50% are protein bound (90% to albumin)

Corrected calcium:
For every 1g/dl (10gm/L) reduction in albumin, total Ca decreases by 0.2 mmo/L
Peggy 18 months old
18 months old girl with bow legs

Biochemistry

Ca 2.19 mmol/L, Albumin 34g/L

PO₄ 1.4mmol/L

ALP 776 IU/L
Other lab tests

PTH 23.3pmol/L (n= 1.3-6.8)

Vitamin D  15nmol/L

Diagnosis : Vit D deficiency
Peggy - Treatment

<table>
<thead>
<tr>
<th>Date</th>
<th>Ca</th>
<th>PO4</th>
<th>ALP</th>
<th>PTH</th>
</tr>
</thead>
<tbody>
<tr>
<td>16.9.05</td>
<td>2.19</td>
<td>1.4</td>
<td>775</td>
<td>23.3</td>
</tr>
<tr>
<td>9.12.05</td>
<td>2.34</td>
<td>1.8</td>
<td>327</td>
<td>8.7</td>
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<tr>
<td>28.6.06</td>
<td>2.34</td>
<td>1.6</td>
<td>212</td>
<td>5.0</td>
</tr>
</tbody>
</table>

Given  vit D 3000 units daily 4 months , followed by maintenance dose of 400 units daily
Ex Premature 35/52, BW 20kg

Corrected age 2 1/12
Had prolonged jaundice
Breast fed exclusively

LFT
Bilirubin 181mmol/L, conjugated 15mmol/L
ALP 620iu/L, AST 27iu/L, ALT 24 iu/L
Ex Premature baby

ALP 726iu/L, AST 21iu/L, ALT 27iu/L

Calcium 2.6mmol/L  PO₄ 1.4mmol/L

treated as rickets of prem, given elemental phosphate
## Endocrine referral

<table>
<thead>
<tr>
<th>ALP</th>
<th>Ca</th>
<th>PO4</th>
</tr>
</thead>
<tbody>
<tr>
<td>807</td>
<td>2.4</td>
<td>1.3</td>
</tr>
</tbody>
</table>

Further lab tests???

PTH and vit D
Endocrine referral

<table>
<thead>
<tr>
<th>ALP</th>
<th>Ca</th>
<th>PO4</th>
<th>PTH</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.1.12</td>
<td>807</td>
<td>2.4</td>
<td>9.7</td>
</tr>
</tbody>
</table>

Vit D = 13.9nmol/L

Diagnosis: vit D deficiency rickets
Treatment: Vitamin D 1000 units daily for 12 weeks, then 400 units daily
# Response to treatment

<table>
<thead>
<tr>
<th></th>
<th>ALP (IU/L)</th>
<th>Ca (mmol/L)</th>
<th>PO4 (mmol/L)</th>
<th>PTH (pmol/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.1.12</td>
<td>807</td>
<td>2.4</td>
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<tr>
<td>16.5.12</td>
<td>207</td>
<td>2.34</td>
<td>1.8</td>
<td>2.0</td>
</tr>
</tbody>
</table>
Hypocalcaemic rickets

Biochemical profile

Low / normal calcium
Normal / Low Pi
High ALP
Albumin (if low, corrected calcium)
High PTH
25hydroxycholecalciferol (VitD)
Vit D deficiency

Serum  25OH vit D < 50nmol/L (20ng/ml)

*Holick* J Nutrition 1990;170: 1464-1469
*Holick*, NEJM 2007; 357:266-281
*Am J Clin Nutr* 2008;87(S1):1080S-1086S
## Vit D (nmol/L)

<table>
<thead>
<tr>
<th>Deficiency</th>
<th>&lt;50nmol/L</th>
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<tbody>
<tr>
<td>Mild</td>
<td>25-50</td>
</tr>
<tr>
<td>Moderate</td>
<td>12.5-25</td>
</tr>
<tr>
<td>Severe</td>
<td>&lt;12.5</td>
</tr>
<tr>
<td>Normal</td>
<td>50-120</td>
</tr>
</tbody>
</table>
Treatment Vit D deficiency
1500-10,000 units D3 for 3/12

Ergocalciferol (VitD3)
infants <1 month: 1000 units/day
1-12 months: 1500-5000 IU/day
older children
  toddlers, children: 6000 IU/day
  adolescents: 10,000 IU/day

treat for 12 weeks
Stoss therapy

100,000 - 600,000 IU  I/M stat  or  
50,000 weekly 1/M  for  8 weeks

Biochemical Response : 1-2 weeks
Severe long standing Vit D deficiency rickets

Elemental Calcium 40-75mg/kg/day in 3 doses in initial phase to avoid hypocalcaemia secondary to “hungry bone” syndrome esp with stoss therapy

Start with higher dose and wean down to lower end by 2-4 weeks, stopped when PTH and vit D is normalised (2 months)

Calcitriol nec: 20-100ng/kg/day in 2-3 doses till normalisation of calcium
symptomatic hypocalcemia

Ca <1.5mmol/L usually symptomatic, give intraveously calcium

Calcium gluconate: 0.3-0.5ml 10% stat, then 2-5mls/kg/day in qid till calcium is 2.0mmol/L

Followed by 40mg/kg day of elemental calcium
Calcium supplement

Calcium gluconate, 500mg tablet
(40%) 200mg elemental calcium/tab

Calcium lactate, 300mg tablet
(13%) 30mg elemental calcium /tab

Food sources:

Milk: a cup of milk has 300mg elemental calcium (3 cups/ day =1000mg)
dried small fish
cheese

Br J Nutr 2000; 83: 191-196
Response

Ca, Pi normalise within 6-10 days,

PTH in 1-2 months

ALP within 3-6 months
Hypocalcaemic Rickets

Once ALP normal i.e. after 12 weeks

Give Maintainance dose of Vitamin D
Daily requirement Vit D (IU)

Prem infants 200-400
Infants to adolescence 400

Source: SUN

: FOOD  VIT D2 (plant)
Vit D3 (animal: fish oil)

*Paediatrics 2008;122: 1142-1152*
Hypocalcaemic rickets

When Vitamin D level is normal, it could be Vitamin defective Metabolism.
Ineffective Vit D action

hereditary defects of Vit D metabolism
Vit D dependant rickets
(I- α hydroxylase enzyme deficiency)

Vit D resistant rickets
(vitamin D receptor defect)
Rickets due to reduced 25-hydroxylase activity

Liver diseases

Vit D dependant rickets
Vitamin D dependant rickets

Autosomal recessive disorder
Failure of $1\alpha$ hydroxylation

- Low $1, 25$ dihydroxycholecalciferol
- Normal $24$ hydroxy cholecalciferol

Low Calcium, Low PO, high ALP

Hyperparathyroidism
Aminoaciduria

NEJM 1998; 338 : 653-661
Case

3 years 11 months girl

Parents nonconsanganeous

delayed motor milestones from 6 months

History frequent falls and waddling gait

Had a fracture left clavicle

No milk/eggs in diet

treated as nutritional Vit D deficiency from 15 months

calcium lactate 30mg qid

calcidiol 0.3mcg daily
Lab results

Ca  1.7mmol/L  (low)
P04  1.1mmol/L  (low)
ALP  3255IU/L
Albumin  45gm/L
U& E normal except CI  112mmol/L

What other tests would you want?
PTH 78 pmol/L (N = 1.3-6.8)
Urine reducing sugar +ve, P04, aminoaciduria

Diagnosis ???????
Further tests
How do you treat????????
Further lab tests

25 cholecalciferol 48nmo/L
1,25 dihydroxycholecalciferol 15pmol/ml

Final diagnosis
Vit D Dependant Rickets
Normal values

25 OH Vit D: 50-120nmol/L

1,25 (OH)2Vit D:
- infants: 70-360pmol/L
- child: 70-220pmol/L
- adult: 50-120pmol/L
Treatment

Vit D dependant rickets

1. Rocalcitriol  2mcg daily

2. Calcium lactate 500mg tds
Progress

Calcium 2.5mmol/L
PO4 1.6mmol/L
ALP 273IU/L

Rx 2.25mcg rocalcitriol
vit D dependant rickets

initial phase
1. Elemental calcium 50mg/kg/day
2. calcitriol : 10-400ng/kg/day
   ie 1-4mcg/day
   1α calcidiol : 2-8mcg/day

Maintenance phase
1. Calcitriol : 0.5-2mcg/day
2. 1α calcidiol : 1-3mcg/day
<table>
<thead>
<tr>
<th>Vitamin Form</th>
<th>$T_{1/2}$</th>
<th>Duration of Action</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cholecalciferol (vit D3)</td>
<td>$T_{1/2}$</td>
<td>2-3 weeks</td>
</tr>
<tr>
<td>$1\alpha$Calcidiol</td>
<td>$T_{1/2}$</td>
<td>30-35hrs</td>
</tr>
<tr>
<td>Calcitriol</td>
<td>$T_{1/2}$</td>
<td>5-8hours</td>
</tr>
</tbody>
</table>
Vit D resistance rickets

Autosomal recessive

Vit D receptor defect

- Defective ligand binding domain (receptor negative subclass)
- Conformational DNA binding domain (receptor positive) : associated with alopecia

Biochemistry : similar to Vit D dependant rickets BUT

1 25dihydroxycholecalciferol is very high
treatment

Receptor negative: Calcitriol 5-60mcg/day

Alopecic children

calcium infused at high doses (2mmol/kg/day up to 2 years, 2-7g/day thereafter)

*J Clin Invest* 1986; 77:1661-1667
*Clin Endocrinol* 1993;30:229-237
## Should Know

<table>
<thead>
<tr>
<th>Old nomenclature</th>
<th>New nomenclature</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Vit D resistant rickets</td>
<td>hypophosphatemic rickets</td>
</tr>
<tr>
<td>2. Vit D dependant type 1 rickets</td>
<td>Vit D dependant rickets (1α hydroxylase defy)</td>
</tr>
<tr>
<td>3. Vit D dependant type 2 rickets</td>
<td>Vit D resistance rickets (rare, receptor defect)</td>
</tr>
</tbody>
</table>
Phosphorous haemostasis

Daily Pi requirement

Prematurity : 60-140mg/kg/day
Term neonate : 300mg/day
Children : 800mg/day
Adolescents : 1200mg/day
Phosphorous haemostasis

phosphate widely available in foods, efficiently absorbed (65%) in the jejunum even in absence of vit D

Passive, dependant on Ca (high calcium salt in diet inhibit absorption)

Active transport: via sodium–phosphate cotransporter 2 (NPT2b)
stimulated by 1,25 vit D, increases PO4 absorption to 85-90%
Phosphorous haemostasis

Low serum phosphate stimulates synthesis of 1,25 dihydroxycholecalciferol

Daily serum phosphate levels vary as much as 50% per day due to effect on food intake

Cicardian rhythm nadir between 7-10am

Best to take PO4 measurement in the basal fasting state
Phosphorous haemostasis

Control of serum phosphate is determined by rate of renal tubular absorption

Bec intestinal phosphate absorption is highly efficient, urinary excretion is not constant, varies with dietary intake

Na/PO4 co transporter (NPT 2a), (NPT2c) reabsorp PO4,
Renal handling Pi

TRP

Spot Urine PO$_4$, creatinine
Plasma PO$_4$ and creatinine

TRP

\[
1 - \frac{\text{Urine PO}_4 \times \text{Scr}}{\text{U Cr} \times \text{SPO}_4}
\]

N = 0.8-0.9
PTH reduces expression of NPT2, Po4 loss

FGR23 impairs phosphate reabsorption
impairs synthesis of 1,25 (OH)\textit{vit} D
which further worsen the resulting hypophosphatemia

FGR23 acts to lower high serum PO4
Phosphate regulation

- Increased urinary 
- Phosphorus (+)
- Phosphatonin (FGF-23)
- 1,25(OH)₂D
- NPT2a
- NPT2c
- 1-α hydroxylase
- Bone
- Kidney
- PTH
- ↑Urinary P

↑iPi
Phosphatemic rickets
deficiency (Prematurity)
loss from kidneys
hereditary

hypophosphataemic rickets
X-linked (XLH) : PHEX mutation
Autosomal dominant (ADHR) : FGF23
Autosomal recessive with hypercalciuria (HHRH) : SLC34A3
X-linked recessive with hypercalciuria : (HHRH) CLCN5
Phosphataemic rickets

Tumour induced rickets
unregulated & excessive secretion of FRG23
frm benign mesenchymal /mixed connective
tissue tumours, head and neck

Fanconi syndrome
diffuse wastage of \( \text{PO}_4 \), \( \text{BG} \), Protein, aa, \( \text{HCO}_3 \)
many causes
Phosphatemic rickets

Deficient intake
chronic debilitatating illness, anorexia nervosa
malabsorption from gut
large antacid use
congenital atresia or postsurgical short gut
Hypophosphataemic rickets

X-linked dominant, 1: 20,000, Xp22.2-p22.1

Inactivating mutation of PHEX gene (phosphate regulating endopeptidase on X-chromosome) results in excess FGF23, a phosphatonin (Pi transport inhibitor) and downregulate s 1 α vit D hydroxylase enzyme.

Clinical: abn LL deformities, appear after infancy stunted growth
no delay in motor milestones (unlike vit D defy disorders)
frontal bossing, late dentition,
Hypophosphataemic rickets

Prone to tooth decay in 2\textsuperscript{nd} decade, dental abscess

In adults calcification in joint capsule causing joint pain & dec range of motion
Pearls

In the presence of low phosphate
if PTH normal, rickets is due to
hypophosphataemic rickets and not due to
secondary hyperparathyroidism
Pearls

X link Hypophosphatemic rickets (excess FGF23)

Biochemistry

Low $P_0^4$, normal Ca, high ALP
Normal PTH
low 1,25 dihydroxycholecalciferol
Normal values

25 OH Vit D | 50-120nmol/L
1,25 (OH)2Vit D

infants | 70-360pmol/L
child | 70-220pmol/L
adult | 50-120pmol/L
Hypophosphataemic rickets

Treatment

1. Elemental Phosphate 40-90mg/kg/day in 5 doses (1-3g/day)
   (to avoid diarrhoea)

2. Calcitriol 15-20 ng/kg/day, increased sever months to maintenance dose of 30-60ng/kg/day
   (1mcg/ml suspension or 0.25mcg/capsule)
Hypophosphataemic rickets

Aim:
doses adjusted to normalise ALP not Pi
avoid hyperparathyroidism & hypercalciuria
monitor Ca, ALP, PTH, urinary Ca or Uca/cr ratio

24hr urinary Ca > 4mg/kg/day: hypercalciuria
Calcium creatinine ratio

Fasting & second void sample (wake up, pass urine, drink lots of water, collect second urine)

serum and urine : calcium, creatinine

Calculate

urine calcium : creatinine ratio
Check for hypercalciuria

<table>
<thead>
<tr>
<th>Urine Calcium (mmol/L)</th>
<th>Creatinine (µmol/L) ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uca 9mmol/L x Pcr(µmol/L)/1000</td>
<td>Pca(mmol/L)xUcr(mmol/l)</td>
</tr>
</tbody>
</table>

Normal values for age:
- 0-6mnths: <2.4mmol/mmol
- 7-18mnths: <1.7
- 18mnths-6yrs: <1.2
- 7yrs- adult: <0.7
<table>
<thead>
<tr>
<th>Biochem Abn</th>
<th>Med Adjustment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum Ca high</td>
<td>hold calcitriol till N</td>
</tr>
<tr>
<td>Urine Ca high</td>
<td>dec calcitriol by 25%</td>
</tr>
<tr>
<td>S PTH high</td>
<td>increase calcitriol by 5% or reduce Pi by 25%</td>
</tr>
<tr>
<td>ALP high ,PTH N</td>
<td>increase Pi by 25%</td>
</tr>
</tbody>
</table>
Hereditary Hypophosphatemic rickets with hypercalciuria (HHRH)

A – recessive, chr 9 q34
loss of function
mutation Sodium dependant phosphate transporter (SLC34A3)

Clinically similar to XLH rickets BUT biochem differ
Low PO4 with high calcitriol, hypercalciuria, high Ca

Treatment: only PO4 50mg/kg/day (max-3gm/day) div doses, BUT calcitriol contraindicated
Hypophosphatemic rickets

PTH
Must check urinary calcium excretion
1, 25 di hydroxy cholecalciferol
Fanconi syndrome

Clinical
FTT, polydipsia, polyuria, rickets

Proximal renal tubules
- glucose, aminoacids, phosphate, bicarbonate
- and uric acids
- aminoaciduria, glycosuria, proteinuria
- high Pi, HCO3, high u pH
Fanconi syndrome

Causes

cystinosis, galactosaemia, tyrosinaemia
GSD, Wilson, Lowe syndrome
heavy metals (lead, mercury, cadmium, uranium)
drugs
aminoglycosides
antineoplastic (cisplat, 6MP, ifosfamide)
Fanconi syndrome

Biochemistry

Low PO4
Normal Ca, , low or normal calcitriol ,
High ALP
Low HCo3, K, Na,
Treatment

1. Correct acidosis

2. PO4 and KCl replacement
Shol’s solution

Each packet of powder contains

Na citrate 10%
Citric acid 2%

Each 10ml solution contains

10mmol Na
6.5mmol of citrate

Stable for a week
Schol ‘s solution

Metabolic acidosis (in 3 doses/day)
Infant and children = 2-3mmol/kg /day of Na

Older children = 5-15mls /dose tds, max 50mls

Adult = 15-30mls /dose, tds, max 90-150mls
Potassium dihydrogen PO4 solution (KH2PO4)

Each 10 mls contain

Na = 9.7 mmol
PO4 = 5.4 mmol/L
elemental phosphate 166 mg
ie 16.6 mg/1ml solution
Summary

Nutritional esp vit D rickets is common preventable if adequate calcium, phosphate, vit D given to high risk groups

- prematurity
- exclusive breast fed infants
- disorders of Gut/ Liver/Kidney

In hypocalcaemic rickets, calcium is not always low, it can be normal with low phosphate depending on stage of disease because of secondary hyperparathyroidism without PTH value, can mistake it as hypocalcaemic rickets or RTA
Summary

Initial biochemistry

- CA/PO/ALP/Albumin/renal profile

- **PTH** is mandatory, it is high
- Vit D,
- Urine
  - Ca, PO4, Creatinine,
  - aa, protein, glucose, pH

In monitoring progress of treatment
- immediate check should be on calcium and phosphate level, by 6-10 days,
- Do not check ALP until 12 weeks
Summary

In hereditary hypocalcaemic rickets
check vit D and 1,25 vit D

In hereditary hypophosphataemic rickets

PTH is normal

Hyphosphataemic rickets needs calcitriol while HHRH calcitriol is contraindicated

Always ensure no hypercalciuria (check calcium creatinine ratio) and check 1,25 vit D (high)

in Treatment do not monitor PO4, instead ALP, S\textsubscript{ca} U\textsubscript{ca}, PTH
Thank You