Case study - Pyloric stenosis

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Case study

Ross is the first born, 6 week old male infant of a Caucasian couple. He is bought to you after the mother has become concerned about his vomiting. Ross weighed 3.5kg at birth and 4.0kg at 3 weeks of age. Subsequently the vomiting started, initially in small amounts and occurring after feeds. Lately the vomits have become larger. There is no bile in the vomit. Over the last few days it seems that baby Ross has vomited most of what he has been feeding. The mother has been breast-feeding but tried to supplement with formula cow’s milk thinking that there was something wrong with her breast milk. The baby has been quite hungry, despite the vomiting and readily accepts another feed. However, over the last couple of days the baby has been less active and feeding poorly.

Self test

Question 1
Before examining Ross what possible causes come to mind?

Select from the following possibilities the ONE most likely cause.

1. Pyloric stenosis
2. Oesophageal atresia
3. Underlying infection (urinary tract, meningitis).
4. Addisonian crisis
5. Gastro-oesophageal reflux

Authors’ Answer is on page 45

Continuing history

On examination, baby Ross appears listless, pulse rate is 180 per min. His weight is 3.7kg. The fontanelle is depressed. The baby feels cold around the hands and feet despite being wrapped appropriately for the weather. You observe some projectile vomiting. You suspect moderate dehydration and volume depletion.

Question 2
Select from the following possible causes the most likely diagnosis

1. Pyloric stenosis
2. Oesophageal atresia
3. Underlying infection
4. Addisonian crisis
5. Gastro-oesophageal reflux

Authors’ Answer is on page 45

Question 3
What is the significance of the depressed fontanelle?

Authors’ Answer is on page 45

Question 4
What is the significance of the cold hands and feet?

Authors’ Answer is on page 45
Continuing history

You order a blood gas examination, serum and urine electrolytes. The results and normal values are as follows:

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Normal values</th>
</tr>
</thead>
<tbody>
<tr>
<td>pH</td>
<td>7.55</td>
<td>(7.35 - 7.45)</td>
</tr>
<tr>
<td>pCO2:</td>
<td>55</td>
<td>(31 - 42)</td>
</tr>
<tr>
<td>HCO3-</td>
<td>46</td>
<td>(20 - 26)</td>
</tr>
<tr>
<td>Sodium</td>
<td>140</td>
<td>(135-145)</td>
</tr>
<tr>
<td>Potassium</td>
<td>3.2</td>
<td>(3.5 - 5.0)</td>
</tr>
<tr>
<td>Chloride</td>
<td>70</td>
<td>(98 - 110)</td>
</tr>
<tr>
<td>Urea</td>
<td>15</td>
<td>(1.5 - 5.0)</td>
</tr>
<tr>
<td>Creatinine</td>
<td>0.08</td>
<td>(0.03 - 0.05)</td>
</tr>
<tr>
<td>Sodium</td>
<td>25</td>
<td>(15-250)</td>
</tr>
<tr>
<td>Chloride</td>
<td>1</td>
<td>(20-250)</td>
</tr>
<tr>
<td>Potassium</td>
<td>50</td>
<td>(25-120)</td>
</tr>
<tr>
<td>pH</td>
<td>8</td>
<td>(4.6-8.0)</td>
</tr>
</tbody>
</table>

**Question 5**
What is the acid base disorder?

Authors’ Answer is on page 45

**Question 6**
Why are the urea and creatinine raised?

Authors’ Answer is on page 45

**Question 7**
Why is there hypokalaemia?

Authors’ Answer is on page 45

**Question 8**
What is the significance of the low serum potassium?

Authors’ Answer is on page 45

**Question 9**
What is the significance of the urine pH?

Authors’ Answer is on page 46

**Question 10**
What is the significance of the low chloride in the urine?

Authors’ Answer is on page 46

Continuing history

The history and examination so far suggests to you that Ross has pyloric stenosis.

**Question 11**
Which area of physical examination will you now do to complete your assessment?

Authors’ Answer is on page 46

Continuing history

On palpation of the abdomen you detect the presence of a pyloric tumour. Looking across the abdomen you observe peristalsis. Ultrasound confirms this. You have now made the diagnosis of pyloric stenosis.

The infant must now be resuscitated in preparation for surgery. Surgery is performed when the electrolytes and acid base abnormalities are corrected. Fluid therapy is instituted to restore body fluids (to stop the secondary hyperaldosteronism) and to correct the alkalosis (to stop new renal bicarbonate formation occurring).

**Question 12**
Predominantly, which two electrolytes must be replenished?

Select ONE option only.

1. Sodium and Chloride
2. Potassium and Chloride
3. Sodium and Potassium
4. Magnesium and Chloride

Authors’ Answer is on page 46

**Question 13**
The oral route for rehydration is preferred, at least initially. Select TRUE or FALSE

Select TRUE or FALSE

Authors’ Answer is on page 46
**Question 14**
What volume of fluids should be returned to the infant over a 24 hour period?

200 ml  
300 ml  
400 ml  
600 ml  

Authors’ Answer is on page 46

**Final history**

An infusion of 5% dextran/0.45% NaCl/40 mmolar KCl is commenced. Ross’s acid-base and body fluid status returns to normal within 24-48 hours of the commencement of fluid replacement. Surgery is successful (Pyloromyotomy) and a biopsy shows pyloric hypertrophy. The child accepts breast milk 2 days later and is soon discharged.

You are sent Ross’s discharge summary and you ask the parents to come in for a follow-up visit. On representation to you in the surgery it is important to ensure that the parents’ questions and anxieties are addressed before discharging baby Ross.
Question 1
Before examining Ross what possible causes come to mind?

Select from the following possibilities the ONE most likely cause.

1. Pyloric stenosis
2. Oesophageal atresia
3. Underlying infection (urinary tract, meningitis).
4. Addisonian crisis
5. Gastro-oesophageal reflux

Author’s answer: Gastro oesophageal reflux.

Feedback: Infants presenting to general practitioners with this scenario are most likely to be caused by gastro-oesophageal reflux.

Question 2
Select from the following possible causes the most likely diagnosis.

1. Pyloric stenosis
2. Oesophageal atresia
3. Underlying infection
4. Addisonian crisis
5. Gastro-oesophageal reflux

Author’s answer: Pyloric Stenosis

Question 3
What is the significance of the depressed fontanelle?

Author’s answer: The fontanelle represents the point of incomplete fusion of the skull bones at the apex of an infant’s head. As such it is continuous with the cerebrospinal fluid. In states of dehydration and consequent reduction in extracellular fluid volume, there will be a commensurate reduction in cerebrospinal fluid and depression of the usually turgid fontanelle.

Question 4
What is the significance of the cold hands and feet?

Author’s answer: In cases of volume depletion there can be a failure of the peripheral circulation resulting in reduced tissue perfusion and hypothermia of the extremities.

Question 5
What is the acid base disorder?

Author’s answer: This infant has a metabolic alkalosis.

Feedback: This is because the pH is raised (hydrogen ion concentration reduced). The expected respiratory response for a metabolic alkalosis is present (the empirical rule that for every mmol/L rise in the plasma bicarbonate concentration from 25, the PaCO2 should rise by 0.7mmHg from 40 is present).

Question 6
Why are the urea and creatinine raised?

Author’s answer: The clinical picture is one of at least moderate extracellular volume depletion with tachycardia and a sunken fontanelle and loss of 10% of body weight. This is associated with reduced renal perfusion and an increase in the urea and creatinine. The urea is increased proportionately more than the creatinine. This is in keeping with a pre-renal injury.

Question 7
Why is there hypokalaemia?

Authors’ answer: The hypokalaemia is secondary to renal loss of potassium (as shown from the urinary electrolytes). The loss of potassium is due to the reduced renal perfusion causing increased release of renin, and sequentially increased angiotensin and aldosterone. This secondary hyperaldosteronism results in potassium secretion into the distal tubule. The potassium secretion is exacerbated by the presence of an alkaline urine.

Question 8
What is the significance of the low serum potassium?

Authors’ answer: Hypokalaemia is one of two potent stimuli to the kidney causing regeneration of bicarbonate (the other is metabolic acidosis). In this case, the generation of new bicarbonate caused by hypokalaemia causes the metabolic alkalosis to persist and is thus deleterious. The correction of the hypokalaemia is thus necessary to get complete resolution of the alkaliotic state.
Question 9
What is the significance of the urine pH8?

Authors’ answer
A urine pH of 8 indicates there is at least 10mmol/L of bicarbonate in the urine. The bicarbonaturia occurs because the renal threshold for reabsorption of bicarbonate (around 25mmol/L normally) is grossly exceeded here. The bicarbonate must be accompanied by a cation (positive ion) for electro-neutrality. The only two cation’s available are sodium and potassium. Thus, even in the presence of extra-cellular depletion there is sodium lost in the urine. As mentioned above, the potassium is high because of the secondary hyperaldosteronism (which acts to increase potassium loss and decrease sodium loss in the urine).

Question 10
What is the significance of the low chloride in the urine?

Author’s answer:
The low urine chloride is an indication that the alkalosis is due to vomiting.

Question 11
Which area of physical examination will you now do to complete your assessment?

Authors’ answer
Examine the abdomen.

Question 12
Predominantly, which two electrolytes must be replenished?
Select ONE option only.

1. Sodium and Chloride
2. Potassium and Chloride
3. Sodium and Potassium
4. Magnesium and Chloride

Authors’ answer
Sodium and Potassium

Feedback:
The principal deficits are in sodium (due to the volume depletion) and potassium (due to the aldosterone-induced kaliuresis). Thus, an infusion of sodium chloride and potassium chloride is most appropriate.

The oral route for rehydration is preferred, at least initially.

Question 13
The oral route for rehydration is preferred, at least initially.

Select TRUE or FALSE

Authors’ Answer
False

Feedback:
The possibility of further vomiting would rule out oral rehydration therapy. Intravenous is preferred.

Question 14
What volume of fluids should be returned to the infant over a 24 hour period?

200 ml
300 ml
400 ml
600 ml

The Authors’ answer is:
400 ml

Feedback:
The volume of fluid returned over the 24 hour period must take into account not only the body fluid deficit upon arrival (300 ml) but also:
- insensible fluid loss (skin, lungs)
- urine loss
- vomiting