Residents Corner: MCQ

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PJKD MCQs Section

Prepare for Exam & Test Your Knowledge!

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MCQ (1)

A 67-year-old man with ESRD due to diabetic nephropathy and hypertension is evaluated for pain and numbness in his right hand. His current hemodialysis access is a right internal jugular tunneled catheter. Previous accesses in the left forearm and left upper arm have failed, and he has no suitable right forearm vessels. Four weeks ago, a right brachiocephalic arteriovenous fistula (AVF) was created. The patient is now experiencing pain, numbness, and weakness of the right hand that are increasing in severity.
On examination, the AVF incision is well healed, and there is a good thrill and bruit; the right radial pulse is palpable but diminished, and the hand is pale and cool. There is a small ulcer on the tip of the index finger. Grip strength and light touch sensation are diminished. Capillary refill is sluggish. Access flow is 500 mL/min.
Which of the following options represents the MOST appropriate management for this patient?

A. refer for urgent banding of AVF
B. refer for urgent arteriography
C. obtain neurophysiologic studies
D. refer for immediate ligation of AVF
E. treat with analgesia

MCQ (2)

MCKD type 2 or Familial Juvenile hyperuricemia nephropathy is caused by mutations in which gene?

A. UMOD
B. NPHP
C. TCF2
D. WNK1
E. TSC1

MCQ (3)

An 84-year-old man with chronic obstructive pulmonary disease (COPD) is admitted to the intensive care unit with toxic megacolon due to *Clostridium difficile* infection. He does not improve with intravenous metronidazole and enteral vancomycin and subsequently undergoes partial colectomy. He is unable to be weaned from the ventilator after surgery, and nutritional supplementation is begun via nasogastric (NG) tube. Since undergoing colectomy, he has had an ileus and minimal stool output.
Due to the ileus, enteral feeding rate has been minimal but 200 mL water every 6 hours has been provided via the NG tube. Daily urine 0utput is 2.8 L.

On physical examination, blood pressure is 122/64 mm Hg, heart rate is 84/min, and weight is 60 kg. He is afebrile. There is no edema, and heart examination is normal. Lung examination reveals an endotracheal tube in place with coarse breath sounds. Abdominal examination is significant for a midline abdominal incision healing normally.

Laboratory data show the following:

	Result	Reference Range
Sodium	149 mEq/L	136 – 145
Potassium	4.1 mEq/L	3.5 – 5.0

Total CO_2	18 mEq/L	23 – 30
Blood urea nitrogen	56 mg/dL	8 – 20
Creatinine	1.2 mg/dL	0.7 – 1.3
Urine		
Sodium	62 mEq/L	Varies with intake
Potassium	43 mEq/L	Varies with intake
Chloride	145 mEq/L	Varies with intake
Osmolality	480 mOsm/kg	Varies with intake

Which one of the following MOST closely approximates the daily electrolyte-free water balance?

A. Negative 0.8L daily water loss
B. 1.4 L daily water gain
C. Negative 2 L daily water loss
D. No gain or loss

MCQ (4)

A 75-year-old woman with a history of osteoarthritis and hypertension presents to the emergency department with lethargy and a 2-day history of diarrhea and vomiting. Her home medications include aspirin 81 mg daily and hydrochlorothiazide 25 mg daily. On her arrival at the emergency department, blood is drawn for analysis, and she is infused with 1 L of 0.9% saline.
Her BP is 92/54 mm Hg and heart rate 110/min. Examination reveals an older woman in no distress with dry mucous membranes and reduced skin turgor. She is confused and slow to respond to directions but is without focal motor or sensory abnormalities on neurologic examination.

Laboratory data include the following:

	Result	Reference Range
Serum		
Sodium	114 mEq/L	136 – 145
Potassium	3.8 mEq/L	3.5 – 5.0
Chloride	80 mEq/L	98 – 106
Blood urea nitrogen	26 mg/dL	8 – 20
Creatinine	1.4 mg/dL	0.5 – 1.1
Glucose	92 mg/dL	70 – 99 (fasting)
Osmolality	250 mOsm/kg	275 – 295
Urine		
Sodium	<20 mEq/L	Varies
Osmolality	550 mOsm/kg	38 – 1400

In addition to discontinuing hydrochlorothiazide, which of the following is the MOST appropriate next step in management?

A. 3% saline at 30 ml per hour
B. 3% saline at 30 ml per hour with 2 ug desmopressin IV
C. Water restriction to 1 L and 0.9% NS at 200 ml/hr
D. NS at 200 ml per hour with desmopressin 2 ug IV
E. Water restriction to 1 L and salt tablet 1 g TDS

MCQ (5)

A 17-year-old boy presents for evaluation of a kidney stone. He developed left flank pain and hematuria 12 weeks previously. Kidney ultrasound showed bilateral nephrocalcinosis and a 4-mm obstructing stone in the left ureter, which passed spontaneously and was found

to be composed of 40% calcium oxalate and 60% calcium phosphate. His maternal uncle has a history of frequent kidney stones. Vital signs, body mass index, and physical examination are normal.
Laboratory data include the following:

	Result	Reference Range
Sodium	140 mEq/L	136 – 145
Potassium	3.3 mEq/L	3.5 – 5.0
Chloride	111 mEq/L	98 – 106
Bicarbonate	19 mEq/L	23 – 28
Creatinine	1.5 mg/dL	0.7 – 1.3
Calcium	9.4 mg/dL	8.6 – 10.2
Phosphorus	2.0 mg/dL	3.0 – 4.5
Parathyroid hormone, intact	20 pg/mL	10 – 65
Alkaline phosphatase	180 U/L	30 – 120
Uric acid	6 mg/dL	3.0 – 7.0
25-OH vitamin D	54 ng/mL	30 – 60
1,25-$(OH)_2$ vitamin D	28 pg/mL	15 – 60
Urine		
Protein-to-creatinine ratio	1.2 mg/mg	<0.2
Albumin-to-creatinine ratio	200 mg/g	<30

A 24-hour urine collection is pending.

Based on available data, which is the MOST likely diagnosis?

A. Distal RTA
B. Sarcoidosis
C. Hypevitaminosis D
D. Hyperparathyroidism
E. Dent disease

Answer Key

MCQ (1): Answer (B)

Explanation:

The patient has signs and symptoms of distal ischemia which require urgent evaluation with arteriography

Placement of an arteriovenous (AV) access creates a shunt that diverts blood flow from the distal extremity. Usually this diversion of blood flow is well tolerated, and the "steal" is insufficient to produce distal ischemia. The most common cause of distal ischemia is vascular disease in the native arterial vessels, compromising inflow to the distal limb. Arterial stenosis located either proximal or distal to the AVF can result in critical ischemia of the distal limb. This is called distal hypo perfusion ischemia syndrome (DHIS). Less common causes of distal ischemia include low cardiac output, hypotension, and/or excessive flow through the AVF. Four percent of upper extremity AVF are complicated by severe DHIS. Upper arm AVF are particularly prone to this complication. Although measurement of access flow and transcutaneous oxygen tension can help, the diagnosis of steal syndrome or DHIS is largely clinical. It is important to recognize signs and symptoms of significant steal and intervene before there is permanent loss of hand function. The precise cause of DHIS will determine the treatment. Thus, the patient should urgently undergo arteriography to evaluate his native arterial vessels. Suitable lesions may be treated with angioplasty. In case of a large anastomosis and high flow fistula, distal revascularization with interval ligation (DRIL) or banding that may be accomplished by a variety of surgical or endovascular techniques can sometimes salvage the access. If no correctable cause of DHIS is identified, the fistula should be ligated.

Although ligation of access will limit further ischemia and potentially restore limb circulation, it would be premature to sacrifice the AVF without first looking for correctable causes. This is of particular importance in this patient without other upper extremity options.

The symptoms of steal should be differentiated from those of carpal tunnel syndrome or other neuropathic process. In cases of diagnostic uncertainty, nerve conduction studies can be obtained. However, such studies are not necessary in this patient with definitive findings on physical examination.

Conservative follow-up of this patient with severe ischemia could result in permanent loss of function or even amputation.

Urgent banding of AVF, while sometimes a treatment for steal, is not indicated without first investigating the cause of ischemia such as arterial stenosis. Banding is usually applied when the AVF flow is considerably higher than the flows measured in this patient.

MCQ (2): Answer (A)

Explanation:

Mutations in the *UMOD* gene cause uromodulin-associated kidney disease. This is also known as Familial gouty nephropathy or *Familial juvenile hyperuricemic nephropathy* or FJHN or MCKD2 or *Medullary cystic kidney disease type 2* or Uromodulin storage disease.

MCQ (3): Answer (A)

Explanation

Electrolyte-free water balance is the difference between water input (intake and metabolic water generation) and water output (renal clearance and insensible losses). Metabolic generation of water results from oxidation of carbohydrate to H_2O and CO_2. Because this amount averages only about 300 mL per day, it can often be disregarded when making clinical estimates. Insensible losses reflect H_2O lost via the skin, respiratory tract, and gastrointestinal tract. Precise measurement of insensible losses is impractical, but can be estimated to be 0.8 – 1.2 L daily under usual circumstances. Tachypnea, fever, burns, wounds and diarrhea can greatly increase insensible losses. Mechanical ventilation with humified air reduces insensible losses.

Renal electrolyte-free water clearance is estimated by the following equation:

$C^{e}_{H2O} = V (1 - [U_{Na} + U_K]/[S_{Na}])$

where C_{eH2O} represents electrolyte-free water clearance, V is the total daily volume of urine, U_{Na} is urine [Na], U_K is urine [K], and S_{Na} is serum [Na].

In this example, the equation becomes:

$C^{e}_{H2O} = 2.8\ L (1 - [62\ mmol/L + 43\ mmol/L]/[149\ mmol/L]) = 0.82\ L$ daily

Intake of electrolyte-free water supplementation = 200 mL × 4 daily or 0.8 L daily.

In this case, it is reasonable to disregard metabolic water generation and to estimate insensible losses to be 0.8 L/day.

Therefore, his net daily electrolyte-free water balance is

0.8 L intake − (0.8 L urinary loss + 0.8 L insensible loss) = −0.8 L.

Note that the use of total urine and serum osmolality, which includes urea and glucose, in place of effective urine and serum osmolality, gives a misleading assessment of free water clearance:

$C_{H2O} = V (1 - [U_{osm}]/[S_{osm}]) = 2.8\ L (1 - [480\ mOsm/kg]/[323\ mOsm/kg] = -1.36\ L$ daily

The calculation for free water clearance using urine osmolality incorrectly predicts that the patient will retain free water.

MCQ (4): Answer (B)

Explanation:

3% saline at 30 mL/h with 2 μg desmopressin IV

Controlled correction of hyponatremia with a combination of 3% saline and IV desmopressin is indicated for this patient with severe hyponatremia of uncertain duration and a risk for over-rapid correction.

This patient presents with hyponatremia of unknown duration, and as a result, she is at risk for osmotic demyelination should she correct too rapidly. Her history—vomiting and diarrhea, thiazide use, low BP, rapid heart rate, poor skin turgor, and low urine sodium—suggests that her hyponatremia is hypovolemic in origin. She is therefore at a high risk for over-rapid correction now that she has received a bolus of normal saline in the emergency room.

In patients with severe hyponatremia, the administration of desmopressin along with 3% saline at the time of presentation has been shown in 2 studies to lead to a predictable rate of increase in serum sodium concentration without over-rapid correction. This proactive approach induces a state similar to the syndrome of inappropriate antidiuretic hormone (SIADH) and prevents the water diuresis seen in patients with hypovolemic hyponatremia after volume resuscitation. Desmopressin should be given every 8 hours until the serum sodium concentration is >125 mEq/L. The rate of administration of 3% saline can be adjusted to achieve the goal increase in sodium of 6 – 8 mEq/L each 24 hours until the sodium concentration approaches normal. Note that over-rapid correction can occur in the setting of thiazide-associated hyponatremia on withdrawal of the offending agent, even in the absence of IV volume expansion.

Water restriction with normal saline could result in over-rapid correction, whereas desmopressin with normal saline could impair correction of hyponatremia by allowing "desalination" of the fluid administered. Similarly, 3% saline administered alone could cause over-rapid correction. Water restriction with salt tablets would likely be insufficient to correct this patient's profound hypovolemia, and its effect on her sodium concentration would be less predictable.

MCQ (5): Answer (E)

Explanation:

The most likely diagnosis in this patient with nephrocalcinosis, increased creatinine, calcium kidney stone, and predominantly non-albumin proteinuria is Dent disease. The hallmarks of this condition are progressive CKD and low molecular weight proteinuria.

Dent disease is an X-linked disorder also known as X-linked recessive nephrolithiasis, X-linked recessive hypercalciuric hypophosphatemic rickets, and low molecular weight proteinuria with hypercalciuria and nephrocalcinosis. The majority of patients have Dent disease 1, caused by an inactivating mutation in the lysosomal chloride transporter gene located on the X chromosome, *CLCN5*. A minority of patients have Dent disease 2, which is caused by a mutation in the *OCRL1* gene, also on the X chromosome. Lowe oculocerebral renal syndrome is also caused by mutation in the *OCRL1* gene, but renal tubular acidosis, congenital cataracts, and intellectual disability characterize this syndrome. There are other patients with the phenotype of Dent disease but without identified mutations in either of these genes.

The pathophysiology of Dent disease is incompletely understood. The chloride channel defect impairs endosome acidification and trafficking, whereas the *OCRL1* gene encodes a phosphatase that also influences endosome trafficking. Failure of the endosome to properly degrade normally filtered low molecular weight proteins leads to the appearance of these proteins in the urine. Retinol binding protein and β-2 microglobulin are often chosen as index proteins for diagnosis. In this case, low molecular weight proteinuria is implied by proteinuria without albuminuria.

There is evidence that the hypercalciuria in Dent disease relates, at least in part, to enhanced intestinal absorption from increased production of 1,25-$(OH)_2$ vitamin D (calcitriol) by the kidney. This may be due to activation of apical membrane parathyroid hormone (PTH) receptors in the late proximal tubule by PTH that has escaped degradation more proximally in the nephron. This paradigm is supported by findings of elevated or high-normal calcitriol, hypophosphatemia, and hyperphosphaturia that are commonly observed in Dent disease. PTH levels are usually low because of negative feedback from high calcitriol. Hyperparathyroidism is ruled out by the relatively low PTH level.

Although elevation of calcitriol can be seen in this diagnosis, this finding is not specific to Dent disease. Elevated calcitriol can also be seen in granulomatous conditions, including sarcoidosis. However, sarcoidosis would not explain low molecular weight proteinuria or hypophosphatemia.

A diagnosis of hypervitaminosis D is not supported by the normal 25-OH vitamin D and phosphorus. Distal renal tubular acidosis would be consistent with the observed electrolyte abnormalities, and may be associated with calcium phosphate stones and nephrocalcinosis, but would not explain proteinuria or hypophosphatemia

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